

2. **Dilation Only:**

- Slight or transient dilation of coronary arteries without aneurysm formation.

3. **Small Aneurysms:**

- Aneurysms with an internal diameter less than 5 mm in children.

4. **Medium Aneurysms:**

- Aneurysms with an internal diameter of 5 mm to 8 mm.

5. **Large or Giant Aneurysms:**

- Aneurysms with an internal diameter greater than 8 mm.
- These pose the highest risk for thrombosis, stenosis, and myocardial infarction.

Aneurysms in K.D:

By absolute dimensions

- Small (<4 Mm Internal Diameter [ID]),
- Medium (>4 To <8 Mm ID), Or
- Giant (>8 Mm ID)

The AHA z -score classification system; z -score-based system (adjusts the coronary dimension for BSA) is as follows:

- No involvement: always < 2 z
- Dilation only: 2 to <2.5 z ;

Or if

Initially <2, a decrease in z score during follow-up >1 z

- Small aneurysm: >2.5 to <5 z
- Medium aneurysm: >5 to <10 z, and absolute dimension <8 mm
- Large or giant aneurysm: >10 z, or absolute dimension >8 mm

Initial Echocardiography is considered positive for purposes evaluation if any of 3 conditions are met:

1. Coronary artery aneurysm is observed; or

2. Z score of left anterior descending coronary artery or right coronary artery >2.5 ;

3. 3 or more other suggestive features exist, including decreased left ventricular function, mitral regurgitation, pericardial effusion, or Z scores in left anterior descending coronary artery or right coronary artery of 2 to 2.5 z.

Note:

If the echocardiogram is positive, treatment should be given within 10 days of fever onset

or

after the tenth day of fever in the presence of clinical and laboratory signs (C-reactive protein [CRP], erythrocyte sedimentation rate [ESR]) of ongoing inflammation

Management of Kawasaki Disease with Coronary Abnormalities

Acute Phase Management

1. Intravenous Immunoglobulin (IVIG):

- Single high-dose IVIG (2 g/kg) is the mainstay of treatment, ideally administered within the first 10 days of illness.
- Reduces the incidence of coronary artery abnormalities.

2. Aspirin Therapy:

- High-dose aspirin (80-100 mg/kg/day) during the acute febrile phase, followed by low-dose aspirin (3-5 mg/kg/day) once fever resolves.
- Continued until platelet count normalizes and no evidence of coronary changes.

Long-term Management and Follow-up

1. Risk Stratification:

- Based on the severity of coronary involvement.
- Regular follow-up with echocardiography to monitor coronary artery status.

2. Anticoagulation Therapy:

- For children with giant aneurysms, anticoagulation with warfarin or low molecular weight heparin, in addition to aspirin, is recommended.
- The goal is to prevent thrombus formation.

3. Beta-Blockers and Statins:

- Considered in cases with significant coronary involvement to reduce myocardial oxygen demand and for atherosclerosis prevention.

4. **Activity Restrictions:**

- May be necessary for children with significant CAAs, especially those with giant aneurysms.

5. **Cardiology Consultation:**

- Essential for ongoing management and decision-making regarding activity level, anticoagulation, and follow-up.

6. **Interventional Procedures:**

- In cases of severe stenosis or obstruction, procedures like angioplasty, stenting, or coronary artery bypass grafting (CABG) may be considered.

Prognosis

- Almost all morbidity and mortality in KD occur in patients with large or giant coronary artery aneurysms, which can be detected and monitored through echocardiography.

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5. Fetal Therapy

Types of Fetal Therapy

1. **Medical Management:**

- Administration of medications to the fetus via the mother.
- Used for conditions like arrhythmias or to enhance lung maturity in cases of anticipated preterm birth.

2. **Minimally Invasive Procedures:**

- Fetoscopic laser photocoagulation for twin-twin transfusion syndrome (TTTS).
- Percutaneous umbilical blood sampling and intrauterine transfusion for conditions like fetal anemia.

3. **Open Fetal Surgery:**

- For conditions like myelomeningocele (spina bifida), congenital diaphragmatic hernia, and some severe forms of congenital heart disease.
- Involves a maternal laparotomy and uterine incision to access the fetus.

4. **Ex Utero Intrapartum Treatment (EXIT):**

- Performed at the time of delivery to secure the airway in fetuses with airway obstruction.

Indications for Fetal Therapy

1. **Congenital Diaphragmatic Hernia (CDH):**

- Fetal endoscopic tracheal occlusion (FETO) can be considered in severe cases.

2. **Lower Urinary Tract Obstruction (LUTO):**

- Vesicoamniotic shunting may be indicated.

3. **Twin-Twin Transfusion Syndrome (TTTS):**

- Fetoscopic laser photocoagulation of the communicating vessels.

4. **Myelomeningocele:**

- Prenatal repair has been shown to improve outcomes compared to postnatal surgery.

5. **Congenital Heart Disease:**

- Fetal aortic valvuloplasty for critical aortic stenosis to prevent hypoplastic left heart syndrome.

6. **Fetal Anemia:**

- Intrauterine transfusion for conditions like Rh disease or fetal-maternal hemorrhage.

Risks and Considerations

1. **Maternal Risks:**

- Includes risks associated with anesthesia, surgical complications, and potential impact on future pregnancies.

2. **Fetal Risks:**

- Risk of preterm labor, fetal injury, or loss during or after the procedure.

3. **Ethical Considerations:**

- Balancing risks and benefits for both mother and fetus.
- Informed consent and multidisciplinary counseling are essential.

Advances and Future Directions

1. **Gene Therapy:**

- Investigational for conditions like congenital adrenal hyperplasia or hemoglobinopathies.

2. **Stem Cell Therapy:**

- Potential future application for a range of genetic and developmental disorders.

3. **Technological Advances:**

- Improvements in imaging and surgical tools are expanding the scope of treatable conditions.

6. Antenatal diagnosis of congenital malformations

Techniques for Antenatal Diagnosis

1. **Ultrasound:**

- The primary modality for screening and diagnosing fetal anomalies.
- Can detect structural anomalies, growth retardation, and some chromosomal abnormalities.
- Advanced ultrasound, including 3D and 4D ultrasounds, enhances the visualization of fetal anatomy.

2. **Maternal Serum Screening:**

- Blood tests to identify markers associated with an increased risk of chromosomal abnormalities (e.g., Down syndrome) and neural tube defects.

3. **Non-Invasive Prenatal Testing (NIPT):**

- Analyzes cell-free fetal DNA in maternal blood.

- Can detect trisomies 13, 18, and 21, and sex chromosome abnormalities.

4. ***Chorionic Villus Sampling (CVS):***

- Performed between 10-13 weeks gestation.
- Involves sampling of placental tissue for chromosomal and genetic analysis.

5. ***Amniocentesis:***

- Usually performed after 15 weeks of gestation.
- Involves the extraction of amniotic fluid for chromosomal, genetic, and biochemical analyses.

6. ***Fetal Magnetic Resonance Imaging (MRI):***

- Used as an adjunct to ultrasound, especially for neurological, thoracic, and complex anatomical evaluations.

7. ***Fetal Echocardiography:***

- Specialized ultrasound to assess the fetal heart, typically performed at 18-22 weeks of gestation.
- Essential for the diagnosis of congenital heart defects.

Commonly Diagnosed Congenital Malformations

1. ***Neural Tube Defects:***

- Includes spina bifida and anencephaly.
- Often detected by elevated alpha-fetoprotein (AFP) levels and ultrasound.

2. ***Chromosomal Abnormalities:***

- Such as Down syndrome, Edwards syndrome (trisomy 18), and Patau syndrome (trisomy 13).
- Diagnosed through NIPT, CVS, or amniocentesis.

3. ***Congenital Heart Defects:***

- Detected via fetal echocardiography.

4. ***Gastrointestinal Anomalies:***

- Like duodenal atresia, gastroschisis, and omphalocele.

5. *Urinary Tract Anomalies:*

- Including kidney agenesis, hydronephrosis, and bladder exstrophy.

6. *Skeletal Dysplasias:*

- Diagnosed through detailed ultrasound examination.

<i>Disorder Type</i>	<i>Specific Disorders</i>	<i>Diagnostic Methods</i>
<i>Chromosomal Abnormalities</i>	Down Syndrome (Trisomy 21) Edwards Syndrome (Trisomy 18) Patau Syndrome (Trisomy 13) Turner Syndrome Klinefelter Syndrome	Non-Invasive Prenatal Testing (NIPT) Chorionic Villus Sampling (CVS) Amniocentesis
<i>Neural Tube Defects</i>	Spina Bifida Anencephaly Encephalocele	Ultrasound Maternal Serum Alpha-Fetoprotein (AFP) Screening Amniocentesis
<i>Congenital Heart Defects</i>	Atrial Septal Defect (ASD) Ventricular Septal Defect (VSD) Tetralogy of Fallot Transposition of the Great Arteries	Fetal Echocardiography Ultrasound
<i>Gastrointestinal Anomalies</i>	Gastroschisis Omphalocele Duodenal Atresia Esophageal Atresia	Ultrasound Fetal MRI (in selected cases)
<i>Urinary Tract Anomalies</i>	Renal Agenesis Hydronephrosis Bladder Exstrophy	Ultrasound Fetal MRI (for detailed assessment)
<i>Skeletal Dysplasias</i>	Achondroplasia Osteogenesis Imperfecta	Ultrasound Fetal MRI (for complex cases)
<i>Genetic Syndromes</i>	Cystic Fibrosis Sickle Cell Disease Thalassemia	CVS Amniocentesis NIPT (for specific known mutations)
<i>Fetal Infections (TORCH)</i>	Toxoplasmosis Rubella Cytomegalovirus (CMV) Herpes Simplex Virus (HSV)	Ultrasound Amniocentesis (for PCR and culture) Maternal Serology
<i>Hematological Disorders</i>	Hemophilia Thalassemia	CVS Amniocentesis NIPT (if specific mutations are known)
<i>Muscular Disorders</i>	Duchenne Muscular Dystrophy	CVS

	Becker Muscular Dystrophy	Amniocentesis NIPT (for known familial mutations)
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- **Ultrasound** is the most common and first-line diagnostic tool for many of these conditions, particularly for structural anomalies.
- **NIPT** is a non-invasive method that analyzes cell-free fetal DNA in maternal blood, primarily used for chromosomal abnormalities.
- **CVS** and **Amniocentesis** are invasive procedures that provide genetic material from the fetus for detailed chromosomal and DNA analysis.
- **Fetal MRI** is used as an adjunct in complex cases where ultrasound is inconclusive or for detailed anatomical assessment.
- **Maternal Serum Screening** and **Fetal Echocardiography** are specific to certain types of disorders like neural tube defects and congenital heart defects, respectively.
- The choice of diagnostic method depends on the suspected disorder, gestational age, and available resources.

Implications of Antenatal Diagnosis

1. **Informed Decision Making:**
 - Allows parents to make informed decisions regarding the continuation of pregnancy and birth plans.
2. **Perinatal Management:**
 - Facilitates planning for delivery in a center with appropriate neonatal care.
 - Enables immediate intervention post-delivery if necessary.
3. **Fetal Therapy:**
 - Some conditions, like TTTS or certain cardiac anomalies, may be amenable to fetal intervention.
4. **Psychological Impact:**
 - The diagnosis of a fetal anomaly can have significant emotional and psychological effects on the parents.

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7. MDR TB diagnosis and management

Clinical Presentation

- Children with MDR-TB often present with non-specific symptoms such as fever, cough, and weight loss.
- The clinical presentation can vary depending on the site of infection, with extrapulmonary manifestations like lymphadenopathy and meningitis also being common.

Investigations

- **Molecular Diagnostics:** Increased availability of molecular diagnostics like Cartridge Based Nucleic Acid Amplification Test (CBNAAT) has the potential to aid in the diagnosis of MDR-TB in children
- **Drug Sensitivity Testing (DST):** Essential for tailoring the treatment regimen.
- **Imaging:** Chest X-rays and CT scans are often used for diagnosis and monitoring.

Treatment principles:

- **Antiretroviral Treatment (ART):** Treatment success rates are higher in children infected with HIV who received ART during MDR-TB therapy
- **Shorter Regimens:** Shorter MDR-TB disease treatment regimens have made therapy safer and shorter
- **Novel Agents and Repurposed Drugs:** Further developments with novel agents and repurposed drugs should lead to additional improvements

Case Definitions

MDR-TB (Multi-Drug Resistant Tuberculosis)

- Tuberculosis that is resistant to at least two of the most potent first-line anti-TB drugs: Isoniazid (H) and Rifampicin (R).

XDR-TB (Extensively Drug-Resistant Tuberculosis)

- A form of MDR-TB that is also resistant to any fluoroquinolone and at least one of three injectable second-line drugs (amikacin, capreomycin, or kanamycin).

Criteria for diagnosis of “Probable MDR-TB” include children with sign and symptoms of active TB disease who in addition have the following risk factors.

They should be considered as having ‘probable’ MDR-TB and started on MDR-TB treatment, even in the absence of bacteriological confirmation.

They include children who have:

- Close contact with a known case of MDR-TB;
- Close contact with a person who died whilst on TB treatment;
- Close contact with a person who failed TB treatment;
- Non response or Failure of a first-line regimen, recognizing that both bacteriological and Clinical definitions of failure should be used; and
- Previous treatment with second-line medications

Treatment Strategies

MDR-TB

1. **Initial Evaluation:** Comprehensive assessment including drug susceptibility testing (DST) to tailor the drug regimen.
2. **Regimen:** Individualized treatment based on DST results, usually involving second-line drugs like fluoroquinolones, aminoglycosides, and other agents like ethionamide, cycloserine, and para-aminosalicylic acid.
3. **Duration:** Extended treatment duration, often up to 24 months.
4. **Monitoring:** Frequent monitoring for drug toxicity and treatment efficacy, often requiring hospitalization initially.
5. **DOTS-Plus:** Directly observed therapy, similar to DOTS but tailored for MDR-TB, to ensure compliance.

XDR-TB

1. **Initial Evaluation:** More extensive DST, including testing for second-line drugs.
2. **Regimen:** Highly individualized, involving newer drugs like bedaquiline and delamanid, and sometimes repurposed drugs like clofazimine and linezolid.
3. **Duration:** Even longer than MDR-TB, often exceeding 24 months.
4. **Monitoring:** Stringent monitoring for drug toxicity and efficacy, with more frequent adjustments to the regimen.
5. **Specialized Centers:** Often managed in specialized centers with expertise in managing complicated forms of TB.

Challenges

- **Diagnosis:** Children with MDR-TB are challenging to diagnose due to non-specific symptoms and the limitations of diagnostic tests.

- **Treatment:** Managing adverse drug reactions and ensuring adherence to treatment regimens are significant challenges.

Future Directions

- **Host-Directed Therapy:** Research is ongoing to explore the role of host-directed therapy in improving treatment outcomes.
- **Comorbid Conditions:** Special attention is required for managing comorbid conditions like HIV.

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8. DKA management

This management strategy is directed towards correcting hyperglycemia, acidosis, and electrolyte imbalances, while identifying and treating the precipitating cause.

Initial Assessment

1. Clinical Evaluation:

- Assess airway, breathing, and circulation (ABCs).
- Evaluate for signs of dehydration, altered mental status, and shock.
- Identify potential precipitating factors (e.g., infection, non-compliance with insulin).

2. Laboratory Assessment:

- Blood glucose, arterial or venous blood gases, serum electrolytes (including sodium, potassium, chloride, bicarbonate), blood urea nitrogen, creatinine.
- Urinalysis for ketones, serum ketones (beta-hydroxybutyrate), complete blood count, and cultures if infection is suspected.
- ECG to monitor for signs of hyperkalemia or hypokalemia.

Fluid Therapy

1. Resuscitation Phase:

- Initial fluid resuscitation with isotonic saline (0.9% NaCl) to restore intravascular volume.
- Typically, 15-20 mL/kg of isotonic saline over the first 1-2 hours.

2. **Maintenance and Deficit Replacement:**

- After initial resuscitation, switch to 0.45% NaCl.
- Calculate fluid deficit and replace over 24-48 hours.
- Adjust fluid type and rate based on serum sodium, glucose levels, and clinical response.

Insulin Therapy

1. **Continuous IV Insulin Infusion:**

- Start with a low-dose insulin infusion (0.1 units/kg/hour), after initial fluid resuscitation.
- Avoid bolus insulin to reduce the risk of cerebral edema.

2. **Transition to Subcutaneous Insulin:**

- Once DKA resolves (blood glucose <200 mg/dL, serum bicarbonate ≥ 18 mEq/L, pH >7.3, and closure of anion gap), transition to subcutaneous insulin.
- Overlap IV insulin with subcutaneous insulin for 1-2 hours to prevent rebound hyperglycemia.

Electrolyte Management

1. **Potassium:**

- Monitor serum potassium levels closely.
- Start potassium replacement if $K^+ < 5.3$ mEq/L and ensure adequate urine output.
- Adjust potassium replacement based on ongoing losses and serum levels.

2. **Phosphate:**

- Routine phosphate replacement is not recommended unless severe hypophosphatemia occurs.

3. **Sodium:**

- Corrected sodium should be monitored to assess the severity of dehydration and response to fluids.

Acid-Base Management

1. **Bicarbonate Therapy:**

- Indicated only in severe acidosis (pH <6.9).
- Use with caution due to potential risks including hypokalemia, cerebral edema, and paradoxical CNS acidosis.

Monitoring

1. Frequent Monitoring:

- Hourly glucose and potassium during the initial treatment phase.
- Regular monitoring of vital signs, urine output, and mental status.

2. Serial Laboratory Assessments:

- Repeat blood gases, electrolytes, and glucose every 2-4 hours until patient is stable.

Management of Complications

1. Cerebral Edema:

- Monitor for signs (headache, altered mental status, bradycardia, hypertension).
- Immediate treatment with mannitol or hypertonic saline if suspected.

2. Hypoglycemia and Hypokalemia:

- Monitor and treat promptly.

3. Infection:

- Identify and treat any underlying or precipitating infection.

Special Considerations

1. Pediatric Patients:

- Greater risk of cerebral edema.
- Fluid replacement should be more cautious.

2. Pregnant Patients:

- Tighter glucose control to avoid fetal complications.

3. Elderly or Cardiorenal Patients:

- Adjust fluid and electrolyte management considering comorbidities.

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9. Criteria for the Classification of Juvenile Idiopathic Arthritis

Criteria:

- Age at onset: <16 yrs.
- Duration of disease: >6 weeks
- Arthritis- swelling or effusion, or the presence of 2 or more of the following signs in >1 joint:
 - limitation of range of motion,
 - tenderness or pain on motion,
 - increased heat
- Onset type defined by type of articular involvement in the first 6 months after onset:
 - Polyarthritis: >5 inflamed joints
 - Oligoarthritis: <4 inflamed joints
 - Systemic-onset disease: arthritis with rash and a characteristic quotidian fever
 - Exclusion of other forms of juvenile arthritis
- **Epidemiology:** No sex predominance in systemic JIA, but more girls than boys are affected in both oligoarticular (3:1) and polyarticular (5:1) JIA. The peak age at onset varies for different types.
- **Etiology:** The etiology and pathogenesis of JIA are largely unknown. It represents a heterogeneous group of disorders sharing the clinical manifestation of arthritis. Genetic factors play a role, but the genetic component is complex.

Classification and Features of Juvenile Idiopathic Arthritis (JIA)/ ILAR classification of juvenile idiopathic arthritis

Classification of JIA	Definition and Criteria	Clinical Features	Exclusions and Special Notes
Systemic JIA	Arthritis in ≥ 1 joint with fever of at least 2 weeks. Accompanied by ≥ 1 of the following: evanescent erythematous rash, generalized lymph node enlargement,	Fever, rash, lymphadenopathy, hepatosplenomegaly, serositis.	Excludes psoriasis, HLA-B27 positive arthritis after 6th birthday, ankylosing spondylitis, enthesitis-related arthritis, sacroiliitis

	hepatomegaly or splenomegaly, serositis.		with IBD, Reiter syndrome, acute anterior uveitis, IgM RF on at least 2 occasions at least 3 months apart.
Oligoarthritis	Arthritis affecting ≤ 4 joints.	Typically affects large joints like knees; may have uveitis.	-
Polyarthritis	Arthritis affecting ≥ 5 joints.	Affects both small and large joints; may be Rheumatoid Factor positive or negative.	-
Psoriatic Arthritis	Arthritis with psoriasis or a family history of psoriasis.	Skin lesions, dactylitis, and nail pitting may be present.	-
Enthesitis-Related Arthritis (ERA)	Arthritis with enthesitis or arthritis or enthesitis with at least two other features such as sacroiliac joint tenderness, inflammatory lumbosacral pain, or HLA-B27 positivity.	Lower back pain, heel pain, joint pain. May also have acute anterior uveitis.	Excludes rheumatoid factor-positive arthritis and other forms of JIA.
Undifferentiated Arthritis	Arthritis that does not fit into the other categories or fits into more than one of the above categories.	Varies; does not meet criteria for other subtypes or meets criteria for more than one subtype.	-

Management of JIA:

Juvenile idiopathic arthritis (JIA) is the most common chronic rheumatic disease in children. The management of JIA is multifaceted and aims to reduce inflammation, control pain, maintain function, and prevent joint damage.

- **Goals of Management:**

- Achieve disease remission or control with minimal side effects.
- Optimize physical and psychosocial growth and development.
- Minimize joint damage and functional disability.

- **Non-pharmacological Interventions:**

- **Physical and Occupational Therapy:** Essential for maintaining joint mobility, muscle strength, and preventing deformities.
- **Joint Protection:** Techniques and devices to protect joints from damage.
- **Patient and Family Education:** Crucial for understanding the disease, treatment options, and coping mechanisms.

- **Pharmacological Interventions:**

1. Non-steroidal Anti-inflammatory Drugs (NSAIDs):

- Examples: Naproxen, Ibuprofen.
- Primarily used to control pain and inflammation.

2. Disease-Modifying Anti-Rheumatic Drugs (DMARDs):

A. Non-biological DMARDs:

Methotrexate: Often the first-choice DMARD for JIA. Helps in reducing joint inflammation.

Sulfasalazine: Especially beneficial for children with enthesitis-related arthritis.

Leflunomide: An alternative for children who can't tolerate methotrexate.

Hydroxychloroquine: Sometimes used, especially in oligoarticular JIA.

Cyclosporine and Azathioprine: Mainly used for systemic JIA.

Cyclophosphamide and Mycophenolate Mofetil: Used in more severe cases or when other treatments fail.

B. Biological DMARDs:

TNF-alpha Inhibitors: Such as etanercept, adalimumab. Effective for treating various subtypes of JIA.

Interleukin (IL) Blockers: Including tocilizumab (IL-6 receptor blocker) and anakinra (IL-1 receptor blocker). Beneficial for systemic JIA.

Abatacept: Blocks T-cell activation. Useful for polyarticular JIA.

Rituximab: Targets B cells. Reserved for severe cases resistant to other treatments.

3. Glucocorticoids:

- Can be given orally, intravenously, or directly into the joint (intra-articular).
- Used for rapid relief but not as a long-term solution due to side effects.

4. Non-pharmacologic Pain Management:

- Techniques like heat/cold applications, relaxation techniques, and distraction can help manage pain.
- **Regular Monitoring and Follow-up:**
 - **Ophthalmological Follow-up:** Regular eye exams are crucial as uveitis (eye inflammation) can develop in children with JIA.
 - **Regular Blood Tests:** Monitor for side effects of medications and disease activity.
 - **Periodic Imaging:** X-rays, MRIs to evaluate joint damage progression.
- **Surgical Intervention:**
 - In rare instances where significant joint damage has occurred, surgical procedures like joint replacement might be considered.
- **Psychosocial Support:**
 - Addressing the emotional and psychological needs of the child and family is paramount. Access to support groups, counseling, and educational resources can be beneficial.

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10. Ocular manifestation of systemic disease

Systemic Disease Category	Specific Disease	Ocular Manifestations
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Endocrine Disorders	Diabetes Mellitus	Diabetic retinopathy (Non-proliferative and proliferative) Diabetic macular edema Early onset cataract Cranial nerve palsies
	Thyroid Disorders (Graves' Disease)	Exophthalmos (proptosis) Eyelid retraction, lagophthalmos Extraocular muscle involvement Exposure keratopathy
Rheumatologic Disorders	Rheumatoid Arthritis	Keratoconjunctivitis sicca Episcleritis and scleritis Secondary Sjögren's syndrome
	Systemic Lupus Erythematosus (SLE)	Retinal vasculitis Cotton wool spots Optic neuropathy
	Ankylosing Spondylitis	Acute anterior uveitis
Infectious Diseases	HIV/AIDS	HIV retinopathy CMV retinitis Kaposi's sarcoma of the eyelid
	Syphilis	Interstitial keratitis Chorioretinitis Optic atrophy
	Tuberculosis	Choroidal tubercles Uveitis
Cardiovascular Diseases	Hypertension	Hypertensive retinopathy Retinal artery and vein occlusions
	Atherosclerosis	Retinal emboli Ischemic optic neuropathy
Hematologic Disorders	Sickle Cell Disease	Retinal vessel occlusions Proliferative sickle retinopathy Retinal detachment
	Leukemia	Retinal hemorrhages Leukemic infiltrates
Neurological Disorders	Multiple Sclerosis	Optic neuritis Internuclear ophthalmoplegia
	Myasthenia Gravis	Ptosis Ocular muscle weakness
Renal Disorders	Hypertensive Nephropathy	Similar to hypertensive retinopathy
	Chronic Renal Failure	Band keratopathy Calcific conjunctival deposits
Gastrointestinal Disorders	Crohn's Disease and Ulcerative Colitis	Episcleritis Uveitis

<i>Dermatological Disorders</i>	Psoriasis	Uveitis Conjunctivitis
	Rosacea	Rosacea keratitis Chalazion and blepharitis
<i>Pediatric rheumatology Disorders</i>	Juvenile Idiopathic Arthritis	Chronic anterior uveitis
<i>Neonate</i>	Retinopathy of Prematurity	In premature infants, related to oxygen therapy
<i>Oncological Disorders</i>	Paraneoplastic Syndromes	CAR (Cancer-associated retinopathy) MAR (Melanoma-associated retinopathy)
	Lymphoma	Intraocular lymphoma

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11. Hearing loss causes and audiological test

Causes of Hearing Loss in Children

Congenital Causes

- Genetic factors
- Maternal infections during pregnancy (e.g., rubella, CMV)
- Prematurity
- Low birth weight
- Anoxia (lack of oxygen) during birth

Acquired Causes

- Chronic ear infections (Otitis media)
- Meningitis
- Measles
- Encephalitis
- Noise exposure
- Ototoxic medications (e.g., aminoglycosides)
- Head trauma
- Tumors affecting the auditory pathway

Syndromic Conditions

- Waardenburg syndrome
- Usher syndrome
- Pendred syndrome
- Alport syndrome

Environmental Factors

- Exposure to loud noises
- Ototoxic chemicals

Other Medical Conditions

- Hyperbilirubinemia
- Autoimmune diseases
- Mumps
- **Michel's Aplasia:** This is a rare congenital condition where the inner ear is completely undeveloped. It results in profound sensorineural hearing loss and is often diagnosed shortly after birth.
- **Mondini's Aplasia:** This is a congenital malformation of the inner ear, specifically the cochlea. It is associated with sensorineural hearing loss and may also be associated with recurrent meningitis.

Causes of Hearing Loss

Category	Specific Causes
Genetic Factors	Congenital anomalies (e.g., Pendred syndrome, Usher syndrome) Genetic mutations affecting ear structure or function
Infections	Congenital infections (e.g., CMV, rubella) Acquired infections (e.g., meningitis, measles, mumps)
Anatomical Abnormalities	Malformations of the ear canal or middle ear Otosclerosis
Trauma	Head injuries Acoustic trauma (e.g., exposure to loud noise) Barotrauma
Ototoxicity	Medications (e.g., aminoglycosides, cisplatin) Chemicals (e.g., solvents, heavy metals)

Age-Related Changes	Presbycusis (age-related hearing loss)
Other Causes	Tumors (e.g., acoustic neuroma) Autoimmune diseases Meniere's disease Sudden idiopathic hearing loss

Audiological Tests

Test	Description	Purpose
Pure Tone Audiometry	Measures hearing sensitivity across a range of frequencies	Determines the degree and type of hearing loss (conductive, sensorineural, or mixed)
Tympanometry	Assesses the function of the middle ear	Identifies middle ear pathologies (e.g., fluid, otosclerosis, eustachian tube dysfunction)
Speech Audiometry	Evaluates understanding and clarity of speech	Assesses speech recognition and comprehension abilities
Otoacoustic Emissions (OAEs)	Measures sounds generated by the cochlea	Screens for cochlear (inner ear) function, especially in newborns and infants
Auditory Brainstem Response (ABR)	Records brainwave activity in response to sound	Assesses the integrity of the auditory pathway up to the brainstem
Electrocochleography (ECoG)	Measures electrical potentials generated in the inner ear	Useful in diagnosing Meniere's disease and other inner ear disorders
Vestibular Evoked Myogenic Potentials (VEMP)	Assesses the saccule and the inferior vestibular nerve	Helps in diagnosing vestibular disorders associated with hearing loss
Immittance Audiometry	Measures the impedance of the middle ear	Helps in identifying middle ear disorders

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12. Adolescent needs and health problems.

- Adolescence, a critical developmental phase between childhood and adulthood, is marked by rapid physical, psychological, and social changes
- Addressing the unique needs and health problems of adolescents is vital for fostering their growth into healthy adults

- The adolescent period is complex and requires a multifaceted approach to health and well-being
- Healthcare providers must be adept at addressing the diverse and evolving needs of adolescents, promoting healthy lifestyles, and intervening early in health problems
- Schools, families, and communities play a crucial role in supporting adolescents' physical, mental, and social well-being
- Comprehensive health education, including sexual and reproductive health, mental health, and substance abuse prevention, is essential
- Regular health check-ups and screenings are recommended to monitor and manage any emerging health issues
- Creating a supportive and understanding environment is key to helping adolescents navigate this challenging yet formative phase of their lives.

Adolescent Needs

1. Nutritional Needs:

- Increased requirements for growth: protein, calcium, iron, vitamins.
- Balanced diet to prevent obesity and eating disorders.

2. Physical Health Needs:

- Regular physical activity.
- Sleep hygiene: Adequate sleep is crucial for growth and mental health.

3. Mental and Emotional Health:

- Support for stress management and mental health issues.
- Development of coping strategies and resilience.

4. Sexual and Reproductive Health:

- Accurate information about sexual development and reproductive health.
- Access to contraception and safe sex education.

5. Educational and Vocational Needs:

- Guidance for career planning and educational support.
- Skills development for future employment.

6. Social Needs:

- Peer relationships: Building healthy and supportive friendships.

- Family support: Nurturing and stable home environment.

7. Safety Needs:

- Safe environments free from violence and abuse.
- Education about substance abuse and its prevention.

Health Problems in Adolescents

1. Mental Health Disorders:

- Depression, anxiety, eating disorders.
- Substance abuse and addiction.

2. Nutritional Issues:

- Obesity and overweight.
- Eating disorders: Anorexia, bulimia.

3. Sexual and Reproductive Health Issues:

- Teenage pregnancies.
- Sexually transmitted infections (STIs).

4. Substance Abuse:

- Alcohol, tobacco, and drug use.
- Risk-taking behaviors leading to accidents and injuries.

5. Chronic Health Conditions:

- Asthma, diabetes, and other chronic illnesses.
- Management of pre-existing pediatric conditions.

6. Injuries and Violence:

- Accidents (e.g., road traffic accidents).
- Physical and sexual abuse.

7. Skin Conditions:

- Acne and other dermatological issues.

8. Oral Health:

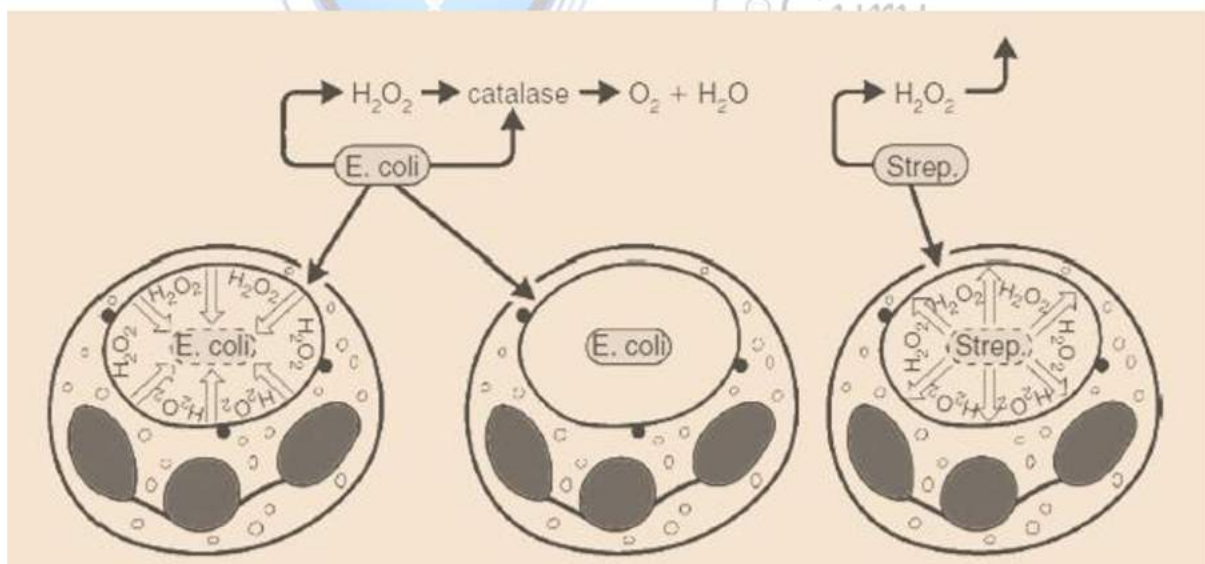
- Dental caries and gum diseases.
- Importance of regular dental check-ups.

13. Chronic granulomatous disease

• **Genetics and Pathogenesis:**

- CGD is a rare immunodeficiency caused by defects in the NADPH oxidase complex.
- It affects phagocytic cells' ability to produce reactive oxygen species (ROS), crucial for killing certain pathogens.
- It's a genetically heterogeneous disease, with mutations in any of the four CGD-related genes (CYBB, CYBA, NCF1, NCF2, and NCF4) causing the disease.
- The CYBB gene, located on the X chromosome, accounts for approximately 65% of cases, making CGD more common in males.
- Mutations in CYBA, NCF1, NCF2, and NCF4 genes are inherited in an autosomal recessive manner.

From the diagram, the pathogenesis of CGD is depicted as follows:



- In normal neutrophils, phagocytosis of pathogens leads to the production of ROS by the NADPH oxidase complex, which is essential for killing bacteria and fungi.
- In CGD, due to defective NADPH oxidase, ROS production is impaired, leading to the survival and proliferation of pathogens within phagocytes.

- The diagram illustrates that catalase-positive organisms like *Streptococcus aureus* can neutralize the low levels of hydrogen peroxide generated by the neutrophils, thereby surviving within the phagocytic cells.
- **Normal Phagocyte:**
 - The left side of the diagram shows a normal phagocyte that has ingested bacteria (*E. coli* and Strep), symbolized by the small red rods and purple cocci within the cell, respectively.
 - Upon ingestion, the phagocyte produces reactive oxygen species (ROS) in the form of hydrogen peroxide (H_2O_2), depicted by the bubbles around the bacteria.
 - The enzyme myeloperoxidase (MPO) uses H_2O_2 to produce hypochlorous acid and other substances to kill the ingested bacteria.
 - Bacteria that produce catalase can convert H_2O_2 to water (H_2O) and oxygen (O_2), which is also illustrated. This is a normal defensive mechanism bacteria use to protect themselves against the oxidative burst of phagocytes.
- **CGD Phagocyte:**
 - The right side of the diagram represents a phagocyte with CGD, which has ingested the same types of bacteria.
 - However, due to the genetic defect in NADPH oxidase, the cell is unable to produce the ROS necessary to effectively kill the bacteria.
 - As a result, the bacteria thrive within the phagocyte, leading to the formation of granulomas, which are depicted as larger clusters within the cell.
 - The lack of ROS production is signified by the absence of bubbles around the bacteria.
- **Clinical Manifestations:**
 - CGD presents with recurrent infections due to the inability of neutrophils to kill certain bacteria and fungi.
 - Common infecting organisms include *Serratia marcescens*, *Burkholderia cepacia*, *Aspergillus* species, and others.
 - Infections can lead to the formation of granulomas, which can obstruct various passages in the body.

- Non-infectious complications include dysregulated inflammation leading to granuloma formation in the gastrointestinal tract and urinary systems, potentially causing obstruction.
- **Laboratory Findings:**
 - The Dihydrorhodamine (DHR) test is used to diagnose CGD by measuring the oxidative burst of neutrophils.
 - Nitroblue tetrazolium (NBT) test is another diagnostic test, but less frequently used due to less sensitivity compared to DHR.
 - Patients may also show abnormal inflammatory markers, such as elevated erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP).
- **Treatment:**
 - Antibiotic and antifungal prophylaxis are key to preventing infections in CGD patients.
 - Interferon-gamma therapy can enhance the immune response in some CGD patients.
 - Corticosteroids and immunomodulatory therapies are used to manage granuloma-related complications.
 - Hematopoietic stem cell transplantation (HSCT) has emerged as a potential cure for CGD, although it comes with significant risks.
- **Genetic Counseling:**
 - Genetic testing for CGD is recommended for at-risk families.
 - Prenatal diagnosis and carrier testing can be performed for families with known mutations.
 - Genetic counseling is crucial to inform families of the inheritance pattern and risks for future offspring.
- **Prognosis:**
 - The clinical course of CGD can vary widely. Early diagnosis and modern treatments have improved the life expectancy of patients.
 - Without proper treatment, serious infections can lead to life-threatening complications.

- Even with prophylactic antibiotics and antifungals, breakthrough infections can occur, necessitating hospitalization and aggressive treatment.
- **Infectious Complications:**
 - The types of infections seen in CGD typically involve organs such as the lungs, lymph nodes, liver, and bones.
 - Pulmonary infections, such as pneumonia, are common, often caused by bacteria like *Staphylococcus aureus* and *Burkholderia cepacia* or fungi like *Aspergillus*.
 - Abscess formation is frequent, and these can be challenging to treat, sometimes requiring surgical intervention.
- **Inflammatory and Autoimmune Phenomena:**
 - CGD patients may experience excessive inflammation even in the absence of infection, leading to conditions like CGD-related colitis.
 - Autoimmune disorders, such as systemic lupus erythematosus (SLE), have been reported in some patients with CGD, although the exact relationship is not fully understood.
- **Diagnostic Imaging and Histopathology:**
 - Imaging studies such as CT scans and MRIs are used to detect infections and granuloma formation.
 - Histopathological examination of biopsied tissues can reveal the presence of granulomas, which are clusters of immune cells that form in response to chronic infection or inflammation.
- **Current Research and Future Directions:**
 - Research is ongoing to better understand the molecular and genetic basis of CGD to improve diagnostics and treatment.
 - Gene therapy trials are exploring the potential to correct the defective genes in hematopoietic stem cells, offering hope for a long-term cure.
- **Patient Management:**
 - Multidisciplinary care is essential, involving immunologists, infectious disease specialists, hematologists, and other healthcare providers.
 - Education of patients and families about the disease, its manifestations, and the importance of adhering to prophylactic treatment is critical.

- Regular follow-ups and monitoring of the patient's health status, immune function, and prompt treatment of infections are crucial components of care.
- **Quality of Life:**
 - The chronic nature of the disease and the need for ongoing treatment can impact the quality of life of patients and their families.
 - Psychological support and counseling may be beneficial to help cope with the challenges posed by the disease.

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14. Primary immunodeficiency management

- **Accurate Diagnosis:**
 - Confirmatory genetic testing for suspected PIDs.
 - Immunological assessments to evaluate specific immune function deficits.
- **Infection Prevention and Control:**
 - Prophylactic antibiotics to prevent bacterial infections.
 - Antifungal and antiviral medications for patients prone to such infections.
 - Immunizations with non-live vaccines according to modified schedules for PID patients.
 - Education on hygiene practices to minimize infection risks.
- **Immunoglobulin Replacement Therapy:**
 - Regular intravenous (IVIG) or subcutaneous (SCIG) immunoglobulin infusions for patients with antibody deficiencies to boost their immune system.
- **Hematopoietic Stem Cell Transplantation (HSCT):**
 - HSCT is considered for severe, combined immunodeficiencies and other select PIDs where it can be curative.
 - Requires a matched donor and carries risks such as graft-versus-host disease.

- **Gene Therapy:**
 - An emerging treatment for certain types of PIDs, where the patient's own cells are genetically modified to correct the immune deficiency.
 - Currently available for a few specific types of PIDs like ADA-SCID (Adenosine Deaminase Deficiency-Severe Combined Immunodeficiency).
- **Enzyme Replacement Therapy:**
 - For specific enzyme deficiencies, like ADA-SCID, where the missing enzyme is provided to the patient.
- **Granulocyte-Colony Stimulating Factor (G-CSF):**
 - G-CSF can be used to stimulate the production of neutrophils in conditions like severe congenital neutropenia.
- **Anti-Inflammatory and Immunomodulatory Therapies:**
 - Corticosteroids and other immunosuppressants can be used to manage autoimmune complications of PIDs.
 - Biologicals like TNF-inhibitors may be employed in certain cases.
- **Treatment of Autoimmunity and Inflammatory Complications:**
 - Autoimmune cytopenias may require immunosuppression, IVIG, or other targeted therapies.
- **Nutritional Support:**
 - Ensuring adequate nutrition, which is vital for immune function and overall health.
 - Supplemental nutrition if gastrointestinal immunodeficiency-related complications are present.
- **Surveillance for Complications:**
 - Regular monitoring for signs of organ damage, malignancy, and other complications associated with PIDs.
 - Routine assessments by a multi-disciplinary team, which may include immunologists, hematologists, infectious disease specialists, and others.
- **Psychosocial Support:**

- Counseling for patients and families to cope with chronic illness.
- Support groups and educational resources for ongoing patient and caregiver support.
- **Special Considerations:**
 - Precautions against live vaccines, which can cause infections in individuals with certain PIDs.
 - Individualized emergency plans for rapid treatment at the onset of infections.

Disorder	Common Features	Management
Antibody Deficiencies		
<i>Common Variable Immunodeficiency</i>	Recurrent bacterial infections, autoimmune phenomena	Immunoglobulin replacement therapy, antibiotics for infections, immunomodulatory therapy
<i>X-Linked Agammaglobulinemia</i>	Absence of B cells, recurrent bacterial infections	Immunoglobulin replacement therapy, antibiotics for infections
<i>Selective IgA Deficiency</i>	Asymptomatic, or recurrent mucosal infections	Antibiotics for infections, avoidance of blood products containing IgA
Combined T and B-Cell Deficiencies		
<i>Severe Combined Immunodeficiency</i>	Severe, recurrent infections, failure to thrive	Hematopoietic stem cell transplantation, prophylactic antibiotics, isolation from infection
<i>Wiskott-Aldrich Syndrome</i>	Eczema, thrombocytopenia, recurrent infections	Immunoglobulin replacement, splenectomy, HSCT
Phagocyte Disorders		
<i>Chronic Granulomatous Disease</i>	Granulomas, recurrent infections, especially fungal	Prophylactic antibiotics and antifungals, interferon-gamma, HSCT
<i>Leukocyte Adhesion Deficiency</i>	Delayed separation of umbilical cord, recurrent skin infections	Antibiotics, bone marrow transplant
Complement Deficiencies		
<i>Hereditary Angioedema</i>	Swelling episodes, abdominal pain	C1 inhibitor concentrate, androgens, antifibrinolytics
<i>C3 Deficiency</i>	Recurrent bacterial infections, immune complex disease	Immunoglobulin replacement, prophylactic antibiotics
T-Cell Deficiencies		
<i>DiGeorge Syndrome</i>	Congenital heart defects, hypocalcemia, T-cell deficiency	Thymus transplant (in select cases), calcium supplementation, immunoglobulin replacement

<i>Chronic Mucocutaneous Candidiasis</i>	Persistent fungal infections of skin and mucous membranes	Antifungal medications, immunomodulatory therapy
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15. Unknown poisoning management

• *Initial Assessment and Stabilization:*

- Assess the child's airway, breathing, and circulation (ABCs). Secure the airway and provide oxygen if needed.
- Check vital signs including pulse, blood pressure, respiratory rate, and temperature.
- Evaluate the need for resuscitation or immediate medical interventions.

• *History and Physical Examination:*

- Attempt to obtain a history of the event from the child, parents, or witnesses.
- Look for any containers, pills, or substances that can give clues to the potential poison.
- Perform a physical exam, noting any odors, burns, skin coloration, or other signs that may indicate the type of poisoning.

• *Decontamination:*

- If the poison was ingested and within an hour of presentation, activated charcoal may be administered to prevent absorption (after ensuring the airway is protected).
- If the poison is on the skin, remove contaminated clothing and wash the skin thoroughly.
- If the poison is in the eyes, rinse the eyes with room temperature water for at least 15 minutes.
- Do not induce vomiting unless advised by a poison control center – Ex AIIMS toxicology helpline. If the following poisonings are suspected one should avoid inducing emesis:

Type of Poison	Reason Vomiting is Contraindicated
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Corrosive Substances Acids and Alkalis	Additional damage to the esophagus and oral mucosa upon vomiting.
Petroleum Products Gasoline, Kerosene, Motor Oil	Risk of aspiration and chemical pneumonitis.
Central Nervous System Depressants - Sedatives, Alcohol	Depressed gag reflex, increased risk of aspiration.
Convulsants - Camphor, Strychnine	Seizures during induced vomiting, aspiration risk.
Hydrocarbons - Paint Thinners, Varnishes	High risk of aspiration into the lungs.
Caustic Substances - Oven and Drain Cleaners	Additional injury to the gastrointestinal tract upon vomiting.
Detergents	Foaming action may lead to aspiration.
Sharp or Solid Objects	Mechanical injury to the gastrointestinal tract.
Pesticides - Organophosphates, Carbamates	Relative contraindication. Potential for seizures or other complications upon vomiting.
Volatile Substances - Inhalants	Increased risk of aspiration; sudden sniffing death syndrome.
Rodenticides	May contain other toxic substances harmful if vomited.

- **Supportive Care:**

- Provide supportive care, including fluids and electrolytes.
- Monitor cardiac function and blood glucose levels.
- Provide symptomatic treatment for seizures, agitation, or other clinical manifestations.

- **Laboratory Tests and Imaging:**

- Order laboratory tests such as complete blood count, electrolytes, blood urea nitrogen, creatinine, liver enzymes, arterial blood gas, and urine analysis.
- Screen for common poisons using toxicology tests.
- Imaging studies (e.g., X-rays) if inhalation or internal trauma is suspected.

- **Consult Poison Control Center:**

- Contact the local poison control center or toxicologist for guidance on further management.
- They can provide information on possible toxins and antidotes.
- **Administration of Antidotes:**
 - If the toxin is known and has a specific antidote, administer it as per the recommended guidelines.
 - Examples include N-acetylcysteine for acetaminophen overdose or atropine for organophosphate poisoning.
- **Continuous Monitoring:**
 - Monitor the child continuously for changes in condition and for the development of any delayed toxic effects.
 - Use telemetry or intensive care monitoring if the child is critically ill.
- **Prevention and Education:**
 - Educate parents and caregivers about poison prevention strategies and keeping potential toxins out of children's reach.
 - Recommend safety latches and proper storage of all hazardous substances.
- **Documentation and Reporting:**
 - Document all findings, treatments, and interventions thoroughly.
 - Report the incident to the appropriate authorities if required by law, especially if there is a suspicion of intentional harm.
- **Psychosocial Support:**
 - Provide support to the child and family, as poisoning events can be traumatic.
 - Consider a referral to social services if there are concerns about the child's home environment.

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16. Nocturnal enuresis

Enuresis refers to the involuntary loss of urine during sleep that occurs at least twice a week in children older than 5 years of age (or the developmental equivalent) for at least 3 months. Usually stops between 5 – 7 years of age.

- ❖ Management of nocturnal enuresis, commonly known as bedwetting, in children involves a combination of behavioral strategies, lifestyle modifications, and medical treatment
- ❖ The approach should be individualized, considering the child's age, emotional state, and the severity of the condition
- ❖ Management of nocturnal enuresis is multifaceted, involving behavioral interventions, lifestyle changes, and possibly medication
- ❖ The approach should be empathetic and supportive, emphasizing that bedwetting is a common pediatric issue and not the child's fault
- ❖ Regular follow-up is important to monitor progress and adjust treatment strategies
- ❖ In cases where standard approaches are ineffective, referral to specialized care may be necessary.

Etiology of Nocturnal Enuresis:

- **Genetic Factors:** A strong family history increases the likelihood of enuresis, suggesting a genetic component.
- **Maturation Delay:** Delay in the development of nocturnal bladder control; the neural pathways responsible for bladder control may mature at a slower rate.
- **Bladder Factors:** A small bladder capacity or overactive bladder muscles can lead to an inability to hold urine produced during the night.
- **Urine Production:** An imbalance in night-time urine production, often due to a lack of normal increase in nocturnal antidiuretic hormone (ADH) levels, which reduces urine production during sleep.
- **Sleep Arousal Difficulties:** Difficulty waking up from sleep when the bladder is full; some children do not awaken easily when their bladder is full.
- **Psychosocial Issues:** Stressful life events or familial stress can be associated with the onset or persistence of bedwetting.
- **Constipation:** Full bowels can place pressure on the bladder, reducing its capacity and exacerbating enuresis.
- **Associated Medical Conditions:** Rarely, enuresis may be related to an underlying medical issue such as a urinary tract infection (UTI), diabetes, or neurological disorders.

Approach- Initial Assessment:**1. Medical History and Physical Examination:**

- Rule out underlying medical conditions (e.g., urinary tract infections, diabetes).
- Assess for associated symptoms (daytime incontinence, constipation, urinary urgency).

2. Psychosocial Assessment:

- Evaluate for any emotional or psychological factors.
- Consider the impact on the child's self-esteem and family dynamics.

3. Voiding Diary:

- Record of fluid intake, urination times, and wetting episodes.
- Helps in understanding patterns and triggers.

Non-Pharmacological Management:**1. Motivational Therapy:**

- Positive reinforcement and encouragement.
- Use of reward systems for dry nights.

2. Bladder Training Exercises:

- Increase bladder capacity and control.
- Scheduled voiding during the day.

3. Fluid Management:

- Avoid excessive fluid intake in the evening.
- Encourage adequate hydration during the day.

4. Bedwetting Alarms:

- Sensor that triggers an alarm at the onset of urination.
- Helps in conditioning the child to wake up or hold urine.

5. Lifestyle Modifications:

- Regular bowel habits to prevent constipation.
- Avoid caffeine and other bladder irritants.

Pharmacological Management:

1. *Desmopressin:*

- Synthetic analog of vasopressin.
- Reduces urine production at night.
- Used for short-term relief (e.g., sleepovers, camps).

2. *Anticholinergic Medications:*

- For children with overactive bladder symptoms.
- Helps increase bladder capacity.

3. *Tricyclic Antidepressants:*

- E.g., Imipramine.
- Used less frequently due to side effects.
- May be considered in resistant cases.

Follow-Up and Support:

1. *Regular Monitoring:*

- Assess response to treatment and adjust as necessary.
- Monitor for any side effects of medications.

2. *Emotional Support:*

- Reassure the child and family that bedwetting is common and treatable.
- Avoid punishment or negative reinforcement.

3. *Referral to Specialists:*

- Consider referral to a pediatric urologist or psychologist if standard treatments are unsuccessful.

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17. POSCO guidelines

- The Protection of Children from Sexual Offences (POCSO) Act, enacted in 2012 in India, is a comprehensive law to provide for the protection of children from the offenses of sexual assault, sexual harassment, and pornography

- It is designed to safeguard the interests and well-being of children, ensuring their safety from sexual crimes
- The POCSO Act is a landmark legislation in India, reflecting the country's commitment to the protection of children from sexual offenses
- It emphasizes the importance of prompt and sensitive handling of cases, ensuring the child's safety and dignity throughout the legal process
- The Act also underscores the role of various stakeholders, including the police, judiciary, medical professionals, and child welfare organizations, in creating a protective environment for children
- Awareness and education about the provisions of the POCSO Act are crucial for its effective implementation and for safeguarding the rights and well-being of children in India.

Key Provisions of the POCSO Act

1. Age of Consent:

- The Act defines a child as any person below the age of 18 years.
- Sexual activities with minors, regardless of consent, are considered offenses.

2. Types of Offenses:

- Penetrative Sexual Assault: Penetration of the penis, any object, or any part of the body into the vagina, mouth, urethra, or anus of a child.
- Aggravated Penetrative Sexual Assault: Includes cases where the abuser is a police officer, public servant, member of the armed forces, etc.
- Sexual Assault: Involving physical contact without penetration.
- Aggravated Sexual Assault: Assault committed by a person in a position of trust or authority.
- Sexual Harassment: Includes non-contact forms of sexual abuse, such as showing pornography to a child.
- Use of Child for Pornographic Purposes: Capturing, disseminating, or using children in pornographic content.

3. Mandatory Reporting:

- It is mandatory to report cases of child sexual abuse. Failure to do so can result in punishment.
- The identity of the child is to be protected at all times during the reporting process.

4. **Child-Friendly Procedures:**

- The Act calls for the establishment of Special Courts to conduct trials in a child-friendly manner.
- The child should not be called repeatedly to testify and should not come in the direct contact with the accused during the trial.

5. **Medical Examination:**

- The child must be given medical care within 24 hours of filing the report.
- Consent of the child or the parent is required for medical examination.

6. **Role of Police and Judiciary:**

- The police are required to file a First Information Report (FIR) in all cases and conduct an investigation.
- The judiciary is required to complete the trial within one year of the offense being reported.

7. **Victim Support and Rehabilitation:**

- The Act provides for the establishment of Child Welfare Committees and Special Juvenile Police Units.
- Psychological support, counseling, and rehabilitation must be provided to the child.

8. **Punishments:**

- The Act prescribes stringent punishments which may include imprisonment and fines, depending on the severity of the offense.

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18. Diphtheria toxins guidelines

- ❖ The management of diphtheria and its toxin requires prompt diagnosis, immediate antitoxin administration, appropriate antibiotic therapy, and supportive care
- ❖ Vaccination remains the cornerstone of prevention
- ❖ Public health measures, including contact tracing and prophylaxis, are essential to control the spread of the disease

- ❖ Healthcare providers should be aware of the potential complications and the special considerations in the management of diphtheria to ensure effective treatment and prevention strategies.

Diagnosis and Initial Management

1. **Clinical Diagnosis:** Its enough to start treatment
 - Symptoms include sore throat, fever, and neck swelling.
 - Presence of a grayish membrane in the throat is characteristic.
2. **Laboratory Confirmation:**
 - Throat swab culture for *Corynebacterium diphtheriae* – Albert stain to be ordered.
 - PCR (Polymerase Chain Reaction) tests for toxin gene detection.
3. **Isolation:**
 - Patients should be isolated to prevent the spread of infection.

Management of Diphtheria Toxin

1. **Antitoxin Administration:**
 - Diphtheria antitoxin neutralizes the circulating toxin. (ADS)
 - Administered as soon as diphtheria is suspected, without waiting for laboratory confirmation.
 - Dosage depends on the severity of the disease.
2. **Antibiotics:**
 - Erythromycin or penicillin is used to eradicate the bacteria.
 - Antibiotics do not neutralize the toxin but prevent further production.
3. **Supportive Care:**
 - Management of airway obstruction.
 - Supportive Treatment of systemic complications like myocarditis and neuropathy.

Prevention and Control

1. **Vaccination:**

- Diphtheria is preventable through vaccination (DPT vaccine).
- Routine childhood immunization and booster doses for adults.

2. **Contact Tracing and Prophylaxis:**

- Identification and surveillance of close contacts.
- Prophylactic antibiotics (erythromycin or penicillin) for close contacts.

3. **Public Health Measures:**

- Reporting cases to health authorities for epidemiological surveillance.
- Health education about the importance of vaccination.

Special Considerations

1. **Allergic Reactions to Antitoxin:**

- Skin testing for hypersensitivity to antitoxin.
- Desensitization protocols if allergic.

2. **Pregnancy and Lactation:**

- Diphtheria antitoxin and antibiotics can be used safely during pregnancy and lactation.

3. **Complications:**

- Myocarditis, neuropathy, and airway obstruction are serious complications.
- Intensive care may be required for severe cases.

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PAPER 4

1. GERD management

- ❖ The management of GERD in infants and children requires a tailored approach, considering the severity of symptoms and the age of the child
- ❖ Lifestyle modifications and dietary changes are first-line interventions
- ❖ Pharmacological treatment is reserved for those who do not respond to these measures or have more severe symptoms
- ❖ Surgical intervention is considered in refractory cases or when complications arise.

Pharmacological Treatment

Proton Pump Inhibitors (PPIs)

- Dosage: Varies by age and weight. For example, omeprazole is often started at 0.7 mg/kg/day for children.
- Common Side Effects: Diarrhea, headache, or upset stomach.
- Duration: Typically prescribed for 4 to 8 weeks but may be extended for long-term treatment.
- Note: PPIs are generally safe and effective but may increase the chance of certain types of infections.

H2 Blockers

- Dosage: For ranitidine, 2-4 mg/kg/day divided into two doses is common.
- Common Side Effects: Abdominal pain, diarrhea, and headache.
- Duration: Short-term use is common, but long-term use is generally avoided due to side effects.

Antacids

- Dosage: Varies by product and age. For example, calcium carbonate may be given at 0.5-1.5 g orally 1-3 times/day.
- Common Side Effects: Diarrhea or constipation.
- Note: Short-term use is recommended due to potential for serious health problems with long-term use.

Non-Pharmacological Treatment

Lifestyle Changes

- Dietary Adjustments: Avoiding spicy, acidic, and fatty foods that trigger symptoms.
- Meal Timing: Avoid eating just before lying down or going to bed.
- Elevated Sleeping Position: Elevating the head of the bed can reduce symptoms.

Postural Adjustments

- For infants, keeping them in an upright position during and after feedings can be beneficial.

Weight Management

- In overweight children, weight loss can significantly improve GERD symptoms.

Surgical Intervention

- Fundoplication: This is the most common surgery for GERD. It involves sewing the top of the stomach around the end of the esophagus to add pressure to the lower esophageal sphincter.
- Risks and Benefits: Surgery is generally considered when medications and lifestyle changes fail. However, children are more likely to develop complications from surgery than from medications.

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2. Recent advances in the management of DMD

- ❖ Duchenne Muscular Dystrophy (DMD) is a severe type of muscular dystrophy characterized by rapid progression of muscle degeneration, leading to muscle weakness and loss
- ❖ Recent advances in the management of DMD focus on genetic, pharmacological, and supportive therapies aimed at slowing the progression of the disease, managing symptoms, and improving quality of life
- ❖ The management of Duchenne Muscular Dystrophy is rapidly evolving, with significant advances in genetic therapies, pharmacological treatments, and supportive care
- ❖ While there is no cure for DMD, these emerging therapies offer hope for slowing disease progression, improving quality of life, and potentially altering the course of the disease

- ❖ Ongoing research and clinical trials continue to expand our understanding and treatment options for DMD, emphasizing the importance of multidisciplinary care and personalized treatment approaches for these patients.

Genetic Therapies

1. Exon Skipping:

- Drugs like Eteplirsen and Golodirsen are designed to skip specific exons in the dystrophin gene, allowing for the production of a partially functional dystrophin protein.
- These therapies are mutation-specific and benefit only a subset of DMD patients.

2. Gene Therapy:

- Research is ongoing in gene replacement therapy, where a functional version of the dystrophin gene is delivered to muscle cells using viral vectors.
- Early clinical trials show promise but are still in the experimental stage.

Pharmacological Advances

1. Corticosteroids:

- Prednisone and deflazacort remain standard treatments to slow muscle degeneration.
- New regimens aim to optimize efficacy while minimizing side effects.

2. Anti-Fibrotic Therapies:

- Investigating drugs that can reduce fibrosis in muscles, potentially preserving muscle function.
- Examples include Halofuginone and Tamoxifen.

3. Cardioprotective Medications:

- ACE inhibitors, beta-blockers, and mineralocorticoid receptor antagonists for managing cardiomyopathy associated with DMD.

4. Novel Anti-Inflammatory Agents:

- Targeting inflammation in dystrophic muscles with drugs like Vamorolone, which may have fewer side effects than traditional steroids.

Supportive and Symptomatic Management

1. Physical Therapy and Rehabilitation:

- Tailored exercise programs to maintain muscle function and prevent contractures.
- Use of assistive devices like braces and wheelchairs.

2. Respiratory Support:

- Non-invasive ventilation for managing respiratory insufficiency.
- Regular monitoring of lung function.

3. Nutritional Support:

- Diet management to prevent obesity, which can exacerbate mobility issues.
- Monitoring and supplementation for nutritional deficiencies.

4. Psychosocial Support:

- Addressing the psychological impact of DMD on patients and families.
- Access to counseling and support groups.

Emerging Research and Trials

1. CRISPR/Cas9 Gene Editing:

- Potential future treatments may involve gene editing to correct mutations in the dystrophin gene.
- Still in early research stages.

2. Stem Cell Therapy:

- Exploring the use of stem cells to regenerate muscle tissue.
- Early-stage research with ongoing clinical trials.

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3. Pediatric covid classification and management

- ❖ The management of COVID-19 in pediatric patients has evolved significantly since the onset of the pandemic

- ❖ The classification and management strategies are tailored to the severity of the disease and the specific needs of children
- ❖ The management of pediatric COVID-19 requires a nuanced approach tailored to the severity of the disease and the individual needs of the child
- ❖ While most children experience mild symptoms, a small percentage may develop severe illness or complications like MIS-C, necessitating intensive care and specialized treatments
- ❖ Ongoing research and clinical experience continue to inform best practices, emphasizing the importance of a flexible, evidence-based approach to the management of COVID-19 in children.

Classification of Pediatric COVID-19

1. **Asymptomatic:**

- Positive for SARS-CoV-2 but no symptoms.

2. **Mild:**

- Upper respiratory tract symptoms like cough, sore throat, nasal congestion, or fever.
- No signs of severe disease.

3. **Moderate:**

- Evidence of lower respiratory disease with symptoms like tachypnea, dyspnea.
- No signs of severe pneumonia.

4. **Severe:**

- Pneumonia with severe symptoms like high fever, severe cough.
- Signs of hypoxia, central cyanosis, or oxygen saturation (SpO₂) <94% on room air.

5. **Critical:**

- Complications such as acute respiratory distress syndrome (ARDS), septic shock, multi-organ failure.

6. **Multisystem Inflammatory Syndrome in Children (MIS-C):**

- A condition where different body parts can become inflamed, including the heart, lungs, kidneys, brain, skin, eyes, or gastrointestinal organs.

Management Strategies

Asymptomatic and Mild Cases

1. *Home Isolation and Monitoring:*

- Most children with asymptomatic or mild COVID-19 can be managed at home.
- Regular monitoring for any signs of disease progression.

2. *Symptomatic Treatment:*

- Hydration, antipyretics for fever and pain.
- No specific antiviral treatment recommended for mild cases.

Moderate Cases

1. *Hospitalization:*

- Consider hospitalization based on clinical judgment, especially for children with underlying conditions.

2. *Supportive Care:*

- Oxygen therapy if SpO₂ is <94%.
- Maintain adequate hydration and nutrition.

3. *Monitoring:*

- Regular monitoring of vital signs, oxygen saturation, and respiratory status.

Severe and Critical Cases

1. *Intensive Care:*

- Admission to a pediatric intensive care unit (PICU) may be necessary.
- Management of ARDS, septic shock, and multi-organ failure.

2. *Advanced Respiratory Support:*

- Non-invasive or invasive mechanical ventilation depending on the severity of respiratory failure.

3. *Pharmacological Treatment:*

- Antiviral agents like Remdesivir (considered in certain cases).

- Immunomodulatory therapy (e.g., corticosteroids, tocilizumab) in the context of severe inflammation or MIS-C.

4. **Antibiotic Therapy:**

- Empirical antibiotics may be considered for possible bacterial co-infection.

MIS-C

1. **Immunomodulatory Therapy:**

- Intravenous immunoglobulin (IVIG).
- Corticosteroids to control inflammation.

2. **Supportive Care:**

- Management of shock, organ dysfunction.
- Cardiac monitoring due to the risk of myocarditis.

COVID Vaccines for prevention:

- Developed to combat the SARS-CoV-2 virus, responsible for the COVID-19 pandemic.
- Multiple platforms used: mRNA, viral vector, protein subunit, and inactivated virus.

Types of COVID-19 Vaccines

- **mRNA Vaccines:** Pfizer-BioNTech, Moderna
- **Viral Vector Vaccines:** AstraZeneca, Johnson & Johnson, Sputnik V
- **Protein Subunit Vaccines:** Novavax
- **Inactivated Virus Vaccines:** Sinopharm, Sinovac-CoronaVac

Mechanism of Action

- **mRNA:** Introduce spike protein mRNA to stimulate immune response.
- **Viral Vector:** Use another virus as a vector to deliver spike protein gene.
- **Protein Subunit:** Use harmless pieces of SARS-CoV-2 to trigger an immune response.
- **Inactivated Virus:** Use a virus that has been killed or inactivated.

Efficacy

- **Pfizer-BioNTech**: ~95% efficacy against symptomatic COVID-19.
- **Moderna**: ~94% efficacy.
- **AstraZeneca**: ~70% efficacy.
- **Johnson & Johnson**: ~66% efficacy.

Recommended Population

- Generally, adults 18 years and older.
- Pfizer-BioNTech approved for adolescents 12-17 in some jurisdictions.

Administration Schedule

- **Pfizer-BioNTech**: Two doses, 21 days apart.
- **Moderna**: Two doses, 28 days apart.
- **AstraZeneca**: Two doses, 4 to 12 weeks apart.
- **Johnson & Johnson**: Single dose.

Adverse Effects

- Common: Pain at injection site, fatigue, headache.
- Rare: Anaphylaxis, myocarditis, blood clotting disorders (AstraZeneca).

Public Health Impact

- Significant reduction in COVID-19 cases, hospitalizations, and deaths.
- Contribution to herd immunity.

Ethical and Policy Considerations

- **Vaccine Equity**: Fair distribution between and within countries.
- **Vaccine Passports**: Ethical implications of requiring proof of vaccination for travel or other activities.

Current Research and Innovations

- Booster doses for waning immunity.
- Vaccines targeting multiple variants.

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4. Malaria vaccines

- ❖ Malaria, caused by Plasmodium parasites transmitted through the bite of infected Anopheles mosquitoes, remains a significant global health challenge
- ❖ The development of malaria vaccines has been a critical area of research, aiming to reduce the incidence and severity of the disease
- ❖ The development of malaria vaccines represents a significant scientific achievement and offers hope for reducing the burden of malaria, particularly in high-risk populations such as children in sub-Saharan Africa
- ❖ While current vaccines like RTS,S provide partial protection, they mark an important step in the global fight against malaria
- ❖ Continued research and development are crucial to improve vaccine efficacy and to address the challenges of production, distribution, and integration with other malaria control strategies.

RTS,S/AS01 (Mosquirix™)

1. *Development and Efficacy:*

- The RTS,S/AS01 vaccine, known as Mosquirix™, is the most advanced malaria vaccine.
- It targets the Plasmodium falciparum parasite, the most deadly malaria parasite globally and the most prevalent in Africa.
- The vaccine has shown to provide partial protection against malaria in young children.

2. *Clinical Trials:*

- Large-scale Phase III clinical trials showed about 36% efficacy in preventing malaria over a four-year period in children who received four doses.
- Efficacy was higher (about 50%) in preventing severe malaria.

3. *WHO Recommendation and Use:*

- In 2015, the World Health Organization (WHO) recommended pilot implementations of the RTS,S vaccine in select areas with moderate to high P. falciparum malaria transmission.
- As of 2021, these pilot programs were underway in Ghana, Kenya, and Malawi.

4. *Vaccination Schedule:*

- The vaccine is administered in four doses: three doses between 5 and 17 months of age and a fourth dose around 18 months later.

Other Malaria Vaccine Candidates

1. R21/Matrix-M:

- A promising candidate showing high efficacy in early trials.
- In Phase IIb trials, it demonstrated over 70% efficacy in one year when given with a booster dose.

2. PfSPZ Vaccine:

- Developed by Sanaria, this vaccine uses sporozoites, the form of the parasite that enters the human liver, to stimulate immunity.
- It has shown promise in early-stage clinical trials.

3. mRNA Malaria Vaccines:

- Following the success of mRNA COVID-19 vaccines, research is underway to develop mRNA vaccines for malaria.
- These vaccines are in the early stages of development.

Challenges and Future Directions

1. Efficacy and Duration of Protection:

- Current malaria vaccines offer partial protection and require multiple doses.
- Research is ongoing to improve efficacy and the duration of protection.

2. Vaccine Production and Distribution:

- Ensuring adequate production and equitable distribution of malaria vaccines, especially in low-resource settings, is crucial.

3. Combination with Other Interventions:

- Vaccines are seen as a complementary tool to existing malaria control measures like bed nets and antimalarial drugs.

4. Continued Research:

- Ongoing research is essential to develop more effective vaccines against different malaria parasites and transmission settings.

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5. All possible radiological and echo findings of RHD

Radiological Findings in Rheumatic Heart Disease

Radiological Feature	Description
Cardiomegaly	Enlargement of the heart silhouette on chest X-ray, indicating chamber dilation or hypertrophy.
Valvular Calcification	Calcification of the mitral and/or aortic valve as seen on chest X-ray or CT scan.
Pulmonary Vascular Changes	Prominent pulmonary artery due to pulmonary hypertension; may also show increased pulmonary vascular markings.
Left Atrial Enlargement	Elevation of the left bronchus and straightening of the left heart border on chest X-ray.
Mitral Valve Changes	Straightening or filling in of the concavity of the left heart border due to mitral valve enlargement.
Aortic Valve Changes	Enlargement of the aortic root or ascending aorta.
Pulmonary Edema	Interstitial or alveolar edema patterns in severe cases, especially with mitral stenosis.

Echocardiographic Findings in Rheumatic Heart Disease

Echocardiographic Feature	Description
Mitral Stenosis	Thickening of mitral valve leaflets, restricted leaflet motion, and narrowed mitral valve orifice.
Mitral Regurgitation	Prolapse or malcoaptation of mitral valve leaflets, leading to backward flow of blood during systole.
Aortic Regurgitation	Incomplete closure of the aortic valve in diastole, leading to diastolic flow reversal in the aorta.
Aortic Stenosis	Thickening and calcification of the aortic valve cusps with restricted movement and narrowed valve orifice.
Left Atrial Dilatation	Enlargement of the left atrium, often secondary to mitral valve disease.
Left Ventricular Hypertrophy	Thickening of the left ventricular walls, usually due to chronic pressure overload.
Chamber Dilation	Dilation of the cardiac chambers, particularly the left atrium and ventricle.
Pulmonary Hypertension	Elevated pulmonary artery pressures, assessed indirectly through tricuspid regurgitation jet velocity.

Subvalvular Changes	Thickening or fusion of the chordae tendineae and papillary muscles, especially in mitral stenosis.
Valvular Vegetations or Thrombi	Rare findings, but may be seen in severe cases with active inflammation or atrial fibrillation.

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6. Fetal surgical therapy

- ❖ Fetal surgical therapy is a rapidly evolving field that offers the potential to significantly improve outcomes for fetuses with certain congenital conditions
- ❖ The decision to proceed with fetal surgery involves careful consideration of the risks and benefits, with a strong emphasis on multidisciplinary care and informed parental consent
- ❖ As technology and surgical techniques continue to advance, the scope of fetal surgery is likely to expand, offering new possibilities for prenatal treatment of complex congenital disorders.

Indications for Fetal Surgery

1. **Spina Bifida (Myelomeningocele):**
 - Repair of open neural tube defects to prevent further neurological damage.
2. **Congenital Diaphragmatic Hernia (CDH):**
 - Repair of diaphragmatic hernia to allow lung development.
3. **Twin-to-Twin Transfusion Syndrome (TTTS):**
 - Laser ablation of placental vessels to correct abnormal blood flow between monochorionic twins.
4. **Lower Urinary Tract Obstruction (LUTO):**
 - Vesicoamniotic shunting to relieve obstruction and prevent renal damage.
5. **Congenital Cystic Adenomatoid Malformation (CCAM):**
 - Resection of lung lesions in severe cases to prevent hydrops and allow lung growth.
6. **Sacroccoccygeal Teratoma (SCT):**

- Resection of large or vascular teratomas to prevent heart failure and hydrops.

Types of Fetal Surgery

1. Open Fetal Surgery:

- Involves a maternal laparotomy and hysterotomy to access the fetus.
- Used for conditions like spina bifida and SCT.

2. Minimally Invasive Fetal Surgery (Fetoscopic Surgery):

- Uses small incisions and fetoscopic techniques.
- Applied in TTTS, LUTO, and some cases of CDH.

3. Percutaneous Procedures:

- Needle-based interventions under ultrasound guidance.
- Includes shunt placements and amnioreduction.

Table: Fetal Surgeries for Various Conditions

Condition	Fetal Surgery	Description
Spina Bifida (Myelomeningocele)	Open Fetal Repair	Surgical closure of the spinal defect before birth to prevent further neurological damage.
Congenital Diaphragmatic Hernia (CDH)	FETO (Fetoscopic Endoluminal Tracheal Occlusion)	Placement of a balloon in the trachea to promote lung growth; removed before birth.
Twin-to-Twin Transfusion Syndrome (TTTS)	Fetoscopic Laser Photocoagulation	Ablation of abnormal vascular connections in the placenta to correct blood flow imbalance between twins.
Lower Urinary Tract Obstruction (LUTO)	Vesicoamniotic Shunting	Placement of a shunt to relieve obstruction and prevent renal damage.
Congenital Cystic Adenomatoid Malformation (CCAM)	Open Fetal Resection or Fetoscopic Laser Ablation	Removal of lung lesions in severe cases to prevent hydrops and allow lung growth.
Sacroccygeal Teratoma (SCT)	Open Fetal Resection	Removal of large or vascular teratomas to prevent heart failure and hydrops.
Congenital Heart Defects	Fetal Cardiac Intervention	Procedures like balloon valvuloplasty or atrial septostomy to treat certain heart defects.

Pulmonary Sequestration	Laser Ablation or Open Resection	Treatment of abnormal lung tissue that is non-functional and separate from the normal lung.
Amniotic Band Syndrome	Fetoscopic Release	Cutting of constricting amniotic bands to prevent limb or digit amputation.
Fetal Anemia (due to Rh Disease)	Intrauterine Transfusion	Transfusion of red blood cells into the fetal circulation.
Twin Reversed Arterial Perfusion (TRAP) Sequence	Radiofrequency Ablation or Coagulation	Ablation of the acardiac twin to improve survival of the pump twin.
Fetal Airway Obstruction (e.g., Congenital High Airway Obstruction Syndrome)	EXIT Procedure (Ex Utero Intrapartum Treatment)	Securing the airway at delivery in cases of anticipated airway obstruction.
Fetal Tumors (e.g., Teratomas, Neuroblastoma)	Open Fetal Resection	Removal of tumors that may compromise the fetus's life or development.

- **Advancements:** New techniques and indications are continually being researched and developed.
- **Multidisciplinary Approach:** These surgeries require a team of specialists, including maternal-fetal medicine physicians, pediatric surgeons, neonatologists, anesthesiologists, and others.
- **Risks and Benefits:** Each surgery carries specific risks and benefits, which must be carefully weighed and discussed with the parents.
- **Postnatal Care:** Many of these conditions require continued care and monitoring after birth.

Risks and Considerations

1. Maternal Risks:

- Includes preterm labor, uterine dehiscence, infection, and anesthesia-related complications.

2. Fetal Risks:

- Potential for preterm birth, intraoperative fetal distress, and procedure-related complications.

3. Long-term Outcomes:

- Vary depending on the condition and the success of the intervention.
- Ongoing follow-up is essential for both the mother and the child.

Ethical and Counseling Considerations

1. **Informed Consent:**

- Parents must be fully informed about the risks, benefits, and potential outcomes.

2. **Multidisciplinary Approach:**

- Involves obstetricians, fetal surgeons, neonatologists, anesthesiologists, and other specialists.

3. **Ethical Considerations:**

- Balancing risks to the fetus and the mother.
- Decisions often involve complex ethical considerations, especially when prognosis is uncertain.

Future Directions

1. **Advancements in Imaging and Surgical Techniques:**

- Improvements in ultrasound, MRI, and fetoscopic tools.

2. **Regenerative Medicine:**

- Potential use of stem cells and tissue engineering in fetal therapy.

3. **Research and Clinical Trials:**

- Ongoing studies to better understand long-term outcomes and refine surgical techniques.

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7. Metabolic acidosis, causes and compensation by body

Classification of Causes of Metabolic Acidosis

- **NAGMA:** Often associated with conditions that lead to a direct loss of bicarbonate or an increase in chloride.
- **HAGMA:** Typically involves the accumulation of organic acids, leading to a higher concentration of unmeasured anions.

Category	Normal Anion Gap Metabolic Acidosis (NAGMA)	High Anion Gap Metabolic Acidosis (HAGMA)
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Gastrointestinal Losses	Diarrhea	-
Renal Disorders	Renal Tubular Acidosis (RTA)	Acute Renal Failure
	Hyperalimentation	Chronic Renal Failure
Drugs and Toxins	Acetazolamide	Methanol
	Spironolactone	Ethylene Glycol
	Ammonium Chloride	Salicylates
Endocrine Disorders	Hyperparathyroidism	Diabetic Ketoacidosis (DKA)
		Alcoholic Ketoacidosis
Ingestions	Chloride Ingestion	Lactic Acidosis
		Aspirin
Respiratory Conditions	Hyperchloremia due to respiratory alkalosis	-
Miscellaneous	Dilutional Acidosis	Sepsis
	Post-Hypocapnia	Starvation
		Rhabdomyolysis

Physiological Compensatory Mechanisms During Metabolic Acidosis

- **Respiratory Compensation:**
 - Hyperventilation to expel CO₂, reducing carbonic acid and increasing pH.
 - Activation of central and peripheral chemoreceptors that sense the decrease in pH.
- **Renal Compensation:**
 - Increased secretion of hydrogen ions in the renal tubules.
 - Enhanced reabsorption of bicarbonate ions.
 - Ammoniogenesis to facilitate hydrogen ion excretion.
- **Buffer Systems:**
 - Activation of intracellular and extracellular buffer systems.

- Phosphate and protein systems in cells neutralize excess hydrogen ions.
- Bicarbonate-carbonic acid system in extracellular fluid.
- **Endocrine Response:**
 - Release of hormones like aldosterone to promote sodium and water reabsorption, indirectly aiding in acid excretion.
 - Parathyroid hormone (PTH) may also play a role in bone buffering of excess hydrogen ions.
- **Shift of Ions Across Cell Membranes:**
 - Intracellular uptake of hydrogen ions in exchange for potassium ions, leading to hyperkalemia.
 - Sodium-hydrogen antiporters also become more active.
- **Metabolic Adjustments:**
 - Mobilization of calcium and magnesium as secondary intracellular buffers.
 - Lactate production may decrease as cells switch to anaerobic metabolism, although this can exacerbate acidosis initially.
- **Cardiovascular Adjustments:**
 - Catecholamine release to maintain cardiac output and tissue perfusion.
 - Peripheral vasoconstriction to maintain blood pressure.
- **Neurological Adjustments:**
 - Activation of survival pathways to protect brain cells from acid-induced damage.
 - Modulation of neurotransmitter release and receptor sensitivity.
- **Hepatic Mechanisms:**
 - Gluconeogenesis may be inhibited to reduce the production of acidic metabolites.
 - Enhanced urea cycle to facilitate ammonia excretion.
- **Muscular System:**
 - Skeletal muscles may increase ammonia production to act as a buffer.

- Reduced muscle contractility due to impaired enzymatic activity in acidic conditions.
- **Immune System Modulation:**
 - Acute phase response with release of cytokines that may have a buffering effect.
 - Modulation of immune cell function to adapt to the acidic environment.

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8. Recent advances in the management of hemophilia

The management of hemophilia is rapidly evolving, with significant strides made in extending the half-life of clotting factors, exploring gene therapy as a potential cure, and developing non-factor therapies

- ❖ Personalized care, improved surgical management, and comprehensive care models are also enhancing the quality of life for patients
- ❖ Ongoing research and global advocacy continue to drive progress in this field, aiming for more accessible and effective treatments for all individuals affected by hemophilia.

1. Extended Half-Life Factor Concentrates

- **Factor VIII and IX Concentrates:** New formulations have been developed that have an extended half-life, reducing the frequency of infusions.
- **Fusion Proteins and Pegylation:** Techniques like fusion to Fc fragments or pegylation extend the half-life of clotting factors.

2. Gene Therapy

- **Potential for a Cure:** Gene therapy holds the promise of a long-term cure for hemophilia by introducing functional copies of the defective gene.
- **Clinical Trials:** Several trials have shown promising results, with some patients achieving sustained normal or near-normal factor levels.

3. Non-Factor Replacement Therapies

- **Emicizumab (Hemlibra):** A bispecific monoclonal antibody that bridges factor IXa and X to restore the function of missing factor VIII. It's used prophylactically in hemophilia A with or without inhibitors.

- **Fitusiran:** An RNA interference therapy that targets antithrombin, increasing thrombin generation and improving clotting.

4. Improved Inhibitor Management

- **Immune Tolerance Induction (ITI) Therapy:** The standard approach for eradicating inhibitors, particularly in hemophilia A.
- **New ITI Strategies:** Research into more effective and less burdensome ITI regimens.

5. Personalized Prophylaxis

- **Tailoring Treatment:** Individualizing prophylaxis based on bleeding phenotype, lifestyle, and pharmacokinetic profiles.
- **Regular Assessment:** Using tools like joint health scores and imaging to guide treatment adjustments.

6. Advances in Surgical Management

- **Safer Surgical Procedures:** Improved perioperative management with factor concentrates and antifibrinolytics.
- **Minimally Invasive Techniques:** Reduced bleeding risk and quicker recovery.

7. Comprehensive Care Models

- **Holistic Approach:** Addressing not just physical but also psychosocial aspects of living with hemophilia.
- **Multidisciplinary Teams:** Involving hematologists, physiotherapists, psychologists, and other specialists.

8. New Diagnostic Tools

- **Genetic Testing:** For precise diagnosis, especially in mild cases or carriers.
- **Point-of-Care Testing:** Potential for home monitoring of factor levels.

9. Global Access and Advocacy

- **Access to Treatment:** Efforts to improve access to advanced therapies in low- and middle-income countries.
- **Patient Advocacy Groups:** Playing a crucial role in education, support, and policy advocacy.

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9. HLH

- ❖ Hemophagocytic lymphohistiocytosis (HLH) is a severe, life-threatening syndrome of excessive immune activation
- ❖ It can be primary (familial) or secondary, associated with infections, malignancies, rheumatologic disorders, or immunodeficiencies
- ❖ The pathophysiology involves uncontrolled and ineffective immune response, leading to tissue damage
- ❖ HLH is a complex and potentially fatal condition requiring prompt recognition and aggressive treatment
- ❖ The management involves a combination of immunochemotherapy, biologic therapies, and HSCT, tailored to the individual patient's condition
- ❖ Ongoing research is focused on improving diagnostic methods, understanding the genetic basis, and developing targeted therapies to improve outcomes in this challenging syndrome.

Pathophysiology

- **Immune Dysregulation:** Central to HLH is the dysregulation of the immune system, particularly involving T cells and macrophages.
- **Cytokine Storm:** Excessive production of pro-inflammatory cytokines, including interferon-gamma, TNF-alpha, IL-6, and IL-18.
- **Genetic Mutations:** In familial HLH, mutations in genes responsible for cytotoxic function of T cells and NK cells are common.

Clinical Features

- **Fever:** Persistent high fever, often unresponsive to standard treatments.
- **Splenomegaly:** Enlargement of the spleen is common.
- **Cytopenias:** Reduction in blood cell counts, including anemia, leukopenia, and thrombocytopenia.
- **Liver Dysfunction:** Elevated liver enzymes, jaundice, and in severe cases, liver failure.
- **Neurological Symptoms:** Irritability, seizures, ataxia, and meningismus.
- **Rash:** Skin rash, often non-specific.

Diagnostic Criteria

- **Clinical and Laboratory Criteria:**

- Fever
 - Splenomegaly
 - Cytopenias
 - Hypertriglyceridemia
 - Hypofibrinogenemia
 - Hemophagocytosis in bone marrow
 - Spleen or lymph nodes
 - Low or absent NK cell activity
 - High levels of ferritin
 - And elevated soluble CD25 (IL-2 receptor alpha).
- **HLH-2004 Protocol:** Widely used for diagnosis, requires five out of eight criteria to be met.
 - **Genetic Testing:** Important for familial HLH.

Treatment

- **Immunotherapy:** The HLH-94/2004 protocol includes dexamethasone, etoposide, and cyclosporine A.
- **Biologic Therapies:** Anti-thymocyte globulin (ATG), rituximab, anakinra (IL-1 receptor antagonist), and tocilizumab (IL-6 receptor antagonist) in refractory cases.
- **Hematopoietic Stem Cell Transplantation (HSCT):** Curative for familial HLH and considered in refractory/relapsed secondary HLH.
- **Supportive Care:** Management of infections, transfusions for cytopenias, and treatment of organ dysfunctions.

Prognosis

- **Variable Outcomes:** Depends on underlying cause, response to treatment, and complications.
- **High Mortality in Severe Cases:** Particularly without prompt and effective treatment.

Research and Future Directions

- **New Therapeutics:** Exploration of novel agents targeting specific pathways in the cytokine storm.

- **Genetic Therapies:** Potential future role for gene therapy in familial HLH.
- **Biomarkers:** Research into biomarkers for early diagnosis and monitoring response to treatment.

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10. Pediatric stroke management

- ❖ Pediatric stroke, though less common than in adults, is a significant cause of morbidity and mortality in children
- ❖ The management of pediatric stroke involves acute intervention, secondary prevention, and rehabilitation
- ❖ The approach differs from adults due to different etiologies, risk factors, and developmental considerations
- ❖ Pediatric stroke management is complex and requires a tailored approach based on the underlying etiology, age, and clinical presentation
- ❖ It involves a combination of acute interventions, secondary prevention, and comprehensive rehabilitation

Acute Management

1. Rapid Assessment and Diagnosis:

- Prompt recognition of symptoms (e.g., weakness, speech difficulties, seizures, headache).
- Neuroimaging: MRI preferred over CT for better sensitivity in detecting ischemic stroke.

2. Acute Intervention:

- **Thrombolysis:** Limited data on safety and efficacy in children; used selectively.
- Drug recommended: Tissue plasminogen activator/ tPA (Alteplase) 0.6 mg/kg (max 50 mg) over 1 hour
- Potential adverse effects: Intracranial hemorrhage; Systemic bleeding; Allergic reactions
- **Mechanical Thrombectomy:** Emerging data support its use in certain cases of large vessel occlusion.

- Management of Increased Intracranial Pressure: Includes head elevation, sedation, and osmotic therapy.

Thrombolysis in Pediatric Stroke

Indication/Cause	Description	Considerations
Acute Ischemic Stroke	Presence of a confirmed acute ischemic stroke with a clear time of onset.	Time window typically within 4.5 hours of symptom onset. MRI or CT confirmation required.
Large Vessel Occlusion	Evidence of large vessel occlusion (e.g., internal carotid artery, middle cerebral artery).	Considered in centers with capability for pediatric neuroimaging and intervention.
No Contraindications	Absence of contraindications such as recent surgery, bleeding disorders, or severe hypertension.	Thorough assessment to rule out risks of bleeding or other complications.
Neurological Deficit	Significant neurological deficit that is disabling, but with potential for recovery.	Assessment of severity and potential for recovery is crucial.
No Hemorrhagic Transformation	No evidence of hemorrhagic transformation on initial imaging.	Hemorrhage risk assessment is critical before administration.
Cardioembolic Stroke	Stroke believed to be due to a cardioembolic source, such as congenital heart disease.	Requires cardiac evaluation and management of underlying cardiac condition.

- **Off-label Use:** Thrombolysis in children is considered off-label, and decisions are often made on a case-by-case basis.
- **Informed Consent:** Due to the off-label nature and potential risks, detailed informed consent is MANDATORY.
- **Post-Thrombolysis Care:** Close monitoring for complications, particularly intracranial hemorrhage, and managing underlying causes.

3. Supportive Care:

- Maintenance of normal blood glucose, oxygenation, and hydration.
- Treatment of seizures if present.
- Blood pressure management: Guidelines less clear than in adults; individualized approach.

Secondary Prevention

1. Identification of Etiology:

- Common causes include arteriopathies, cardiac disorders, sickle cell disease, and prothrombotic states.
- Comprehensive evaluation to determine the underlying cause.

2. **Antithrombotic Therapy:**

- Anticoagulation (e.g., warfarin, heparin) or antiplatelet agents (e.g., aspirin) depending on the etiology.
- Long-term management based on the risk of recurrence and underlying condition.

3. **Management of Risk Factors:**

- Control of modifiable risk factors like obesity, hypertension, and hyperlipidemia.
- Regular follow-up and monitoring for recurrence or complications.

Rehabilitation and Recovery

1. **Multidisciplinary Rehabilitation:**

- Involves physical therapy, occupational therapy, speech therapy, and educational support.
- Early intervention to maximize recovery and adapt to disabilities.

2. **Psychological Support:**

- Addressing the emotional and psychological impact on the child and family.
- Counseling and support groups.

Research and Future Directions

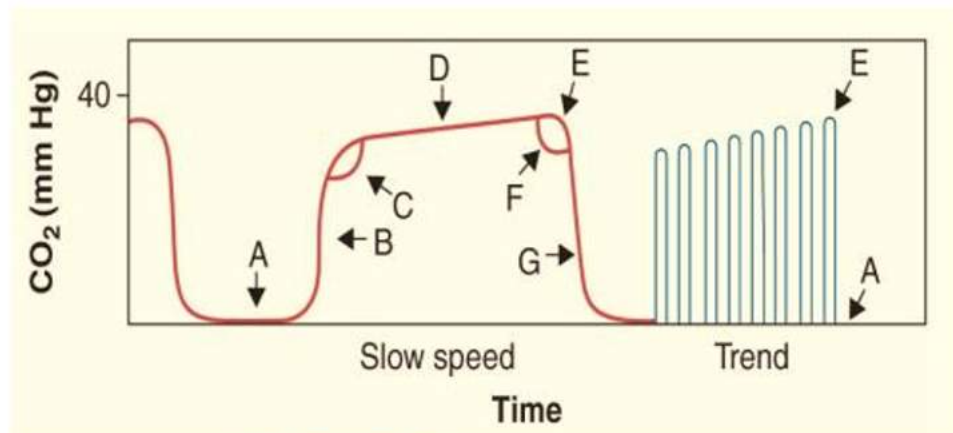
1. **Clinical Trials:** More pediatric-specific research is needed, particularly in acute interventions.
2. **Biomarkers:** Investigating biomarkers for early detection and prognosis.
3. **Neuroprotective Strategies:** Exploring treatments to protect the brain and enhance recovery.

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11. EtCO₂, Measurement, and interpretations

- ❖ EtCO₂ monitoring is not just a tool for respiratory assessment but also has implications in hemodynamic monitoring, especially in critical care settings.
- ❖ It can be used to assess the effectiveness of CPR, guide ventilation in mechanically ventilated patients, and serve as an early warning system for various respiratory and systemic complications.

Normal features of a capnogram:



A - baseline, represents the beginning of expiration and should start at zero.

B - the transitional part of the curve, represents mixing of dead space and alveolar gas.

C - the α angle, represents the change to alveolar gas.

D - the alveolar part of the curve, represents the plateau average alveolar gas concentration.

E - is the end-tidal CO₂ value.

F - the β angle, represents the change to the inspiratory part of the cycle.

G - the inspiration part of the curve, shows a rapid decrease in CO₂ concentration

Increases in $ETCO_2$

Increased pulmonary capillary blood flow
 Increased cardiac output
 Hypoventilation
 Increased CO_2 production
 Sudden release of a tourniquet
 Sodium bicarbonate administration

Decreases in $ETCO_2$

Decreased pulmonary capillary blood flow
 Pulmonary hypertension
 Pulmonary embolus (thrombus or air)
 Decreased cardiac output
 Hyperventilation
 Ventilator circuit leak
 Obstructed endotracheal tube
 Decreased CO_2 production

Absent $ETCO_2$

Esophageal intubation
 Ventilator disconnect

False-positive $ETCO_2$ values - occur when the ETT is in the esophagus (following mouth to-mouth ventilation or ingestion of antacids or carbonated beverages) or when the tip of the ETT is in the pharynx.

False-negative $ETCO_2$ readings (no $ETCO_2$ when the ETT is in trachea) - occur with severe airway obstruction, poor cardiac output, pulmonary emboli, or pulmonary hypertension.

During cardiopulmonary resuscitation, capnography is utilized to assess the quality of chest compressions and provide an early indication of return of spontaneous circulation.

Effective chest compression should produce enough pulmonary blood flow to generate an exhaled CO_2 value of >10 mm Hg.

Time-Based versus Volumetric Capnography

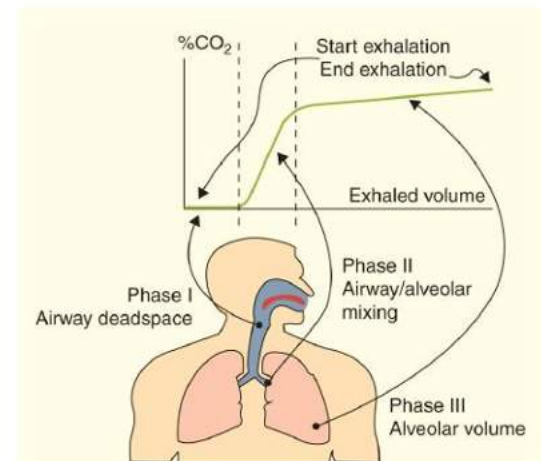
- Time-based capnography is limited to the measurement and display of the respiratory waveform and $ETCO_2$ value

- ETCO₂ can be used to trend changes in PaCO₂ for patients in whom physiologic dead space is not significantly elevated or changing
- The integration of flow and volume graphically displayed is the basis of volumetric capnography
- Volumetric capnogram.

❖ **Phase I** - The initial portion of the volumetric capnogram represents the quantity of CO₂ eliminated from the large airways.

❖ **Phase II** is the transitional zone, which represents ventilation from both small and large airways.

❖ **Phase III** represents CO₂ elimination from the alveoli and, thus, the quantity of gas involved with alveolar ventilation.



- Volumetric capnography provides a measurement of CO₂ production (VCO₂)
- and enables calculation of alveolar minute ventilation and the ratio of volume of dead space- (V_d) to tidal volume (V_T) → V_d/V_T
- V_d/V_T values of <0.5–0.55 and >0.65 - as predictive of extubation success and failure, respectively.

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12. ADHD management

- **Core Symptoms:** ADHD is characterized by a persistent pattern of inattention and/or hyperactivity-impulsivity that interferes with functioning or development.
- **Inattention:** Six or more symptoms of inattention for children up to age 16, or five or more for adolescents 17 and older and adults; symptoms must be present for at least 6 months, and they must be inappropriate for developmental level:

- Often fails to give close attention to details or makes careless mistakes in schoolwork, at work, or with other activities.
- Often has trouble holding attention on tasks or play activities.
- Often does not seem to listen when spoken to directly.
- Often does not follow through on instructions and fails to finish schoolwork, chores, or duties in the workplace.
- Often has trouble organizing tasks and activities.
- **Hyperactivity and Impulsivity:** Six or more symptoms of hyperactivity-impulsivity for children up to age 16, or five or more for adolescents 17 and older and adults; symptoms must be present for at least 6 months to an extent that is disruptive and inappropriate for the person's developmental level:
 - Often fidgets with or taps hands or feet, or squirms in seat.
 - Often leaves seat in situations when remaining seated is expected.
 - Often runs about or climbs in situations where it is not appropriate.
 - Often unable to play or take part in leisure activities quietly.
 - Is often "on the go" acting as if "driven by a motor".
- **Age of Onset:** Several inattentive or hyperactive-impulsive symptoms were present before age 12 years. *(This is recently revised from 7yrs to 12 yrs)*
- **Settings:** Several symptoms are present in two or more settings, such as at home, school, or work; with friends or relatives; in other activities.
- **Impairment:** There is clear evidence that the symptoms interfere with, or reduce the quality of, social, school, or work functioning.
- **Exclusion Criteria:** The symptoms do not happen only during the course of schizophrenia or another psychotic disorder and are not better explained by another mental disorder.
- **Subtypes:** ADHD is categorized into three subtypes—Predominantly Inattentive, Predominantly Hyperactive-Impulsive, and Combined Presentation.
- **Co-morbid Conditions:** Commonly co-occurs with conditions like Oppositional Defiant Disorder, Anxiety Disorders, and Learning Disabilities.
- **Diagnostic Tools:** Standardized rating scales like the Conners' Rating Scales or ADHD Rating Scale-IV are commonly used.

Treatment: Multimodal treatment approach including pharmacotherapy (usually stimulant medications), behavioral therapy, and educational interventions.

- **Pharmacotherapy:**

- **Stimulant Medications:** Methylphenidate, Dexmethylphenidate, Amphetamine, and Dextroamphetamine are first-line treatments.
- **Non-Stimulant Medications:** Atomoxetine, Guanfacine, and Clonidine are alternatives for those who do not respond well to stimulants.

Table on Drugs Used in ADHD Management

Drug Category	Drug Name	Dosage	Side Effects
Stimulants; First-line treatment	Methylphenidate	10-60 mg/day	Insomnia, Decreased Appetite, Weight Loss
	Dexmethylphenidate	5-20 mg/day	Same as Methylphenidate
	Amphetamine	5-40 mg/day	Same as Methylphenidate
	Dextroamphetamine	5-40 mg/day	Same as Methylphenidate
Non-Stimulants (Alternative to stimulants)	Atomoxetine	40-100 mg/day	Fatigue, Nausea, Mood Swings
	Guanfacine	1-4 mg/day	Drowsiness, Fatigue
	Clonidine	0.1-0.3 mg/day	Drowsiness, Dry Mouth

- **Behavioral Therapy:**

- **Cognitive Behavioral Therapy (CBT):** Helps in self-regulation and coping strategies.
- **Parent-Child Interaction Therapy (PCIT):** Focuses on improving parent-child interactions and managing child behavior.
- **Social Skills Training:** For improving social interactions and reducing disruptive behaviors.

- **Educational Interventions:**
 - **Individualized Education Program (IEP):** Tailored educational plan.
 - **Classroom Accommodations:** Extra time for tests, frequent breaks, and a quiet environment for work.
- **Psychoeducation:**
 - Educating the child, family, and teachers about ADHD.
 - Strategies for improving organizational skills, time management, and problem-solving.
- **Nutritional Management:**
 - Omega-3 fatty acid supplements have shown some promise.
 - Monitoring food & additives that may exacerbate symptoms.
- **Physical Activity:**
 - Regular exercise can help in managing symptoms.
- **Mindfulness and Relaxation Techniques:**
 - Mindfulness meditation, yoga, and other relaxation techniques can help in focusing.
- **Co-morbid Conditions:**
 - Treatment for co-occurring disorders like anxiety, depression, or Oppositional Defiant Disorder (ODD) is crucial.
- **Follow-up and Monitoring:**
 - Regular follow-up with healthcare providers for medication management.
 - Monitoring for potential side effects like sleep problems or changes in appetite.
- **Community Support:**
 - Support groups and forums can provide additional resources and emotional support.

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13. CTEV management

- ✚ The treatment plan should be individualized based on the severity of the deformity and the response to treatment.
- ✚ Family education and support are integral parts of the management plan.
- ✚ Lifelong follow-up may be necessary in some cases to monitor for recurrence or complications.

Age Group	Initial Assessment	Treatment Modalities	Follow-up and Monitoring	Additional Notes
Newborn 3 months	Physical examination Radiographic assessment	Ponseti method Serial casting Achilles tenotomy if needed	Weekly casting Monitoring for skin irritation or complications	Early intervention is crucial for optimal outcomes.
3 months 1 year	Re-assessment of deformity Radiographic assessment if needed	Continuation or modification of Ponseti method Surgical intervention if casting fails	Monthly to quarterly visits Monitoring for recurrence	Foot abduction braces are often used after successful casting.
1 year 3 years	Assessment of gait Radiographic assessment	Surgical intervention for residual or recurrent deformities Physical therapy	Quarterly to bi-annual visits Gait analysis	Footwear modification may be needed.
3 years 6 years	Gait analysis Radiographic assessment	Surgical intervention for residual deformities Physical therapy	Bi-annual visits Monitoring for complications of surgery	Social and psychological support for school integration.
6 years and above	Detailed musculoskeletal and functional assessment	Surgical interventions for complications or residual deformities Physical therapy	Annual visits Long-term monitoring for foot and ankle function	May require adaptive devices for mobility.

Initial Assessment

- **Physical Examination:** Assess foot deformity, flexibility, and presence of other congenital anomalies.
- **Family History:** Evaluate for genetic factors, as CTEV can have a hereditary component.
- **Ultrasound or X-Ray:** Imaging is used in atypical cases or older children to assess bone structure.

Non-Surgical Treatment

1. Ponseti Method:

- **Manipulation and Casting:** Begins shortly after birth. Weekly gentle manipulation and casting to gradually correct the foot position.
- **Achilles Tenotomy:** Often required in about 90% of cases to correct equinus deformity.
- **Bracing:** After correction, foot abduction braces are used to maintain correction. Initially full-time, then only during naps and night-time up to 4 years of age.

2. French Functional (Physical Therapy) Method:

- Daily stretching, mobilization, and taping.
- Requires active participation from parents/caregivers.
- Used as an alternative or adjunct to the Ponseti method.

Surgical Treatment

- **Indications:** Reserved for cases where non-surgical methods fail or in older children with neglected CTEV.
- **Types of Surgery:**
 - Soft tissue release (posterior, medial, and sometimes lateral release).
 - Osteotomies for severe deformities or older children.
 - Tendon transfers for dynamic deformities or recurrences.

Post-Treatment Monitoring

- **Regular Follow-Up:** To monitor for recurrence, which is common in CTEV.
- **Physical Examination:** Assess foot alignment and flexibility.
- **Parental Education:** On the importance of brace compliance and monitoring for signs of recurrence.

Complications and Management

- **Recurrence:** Managed with repeat casting, possible surgery.
- **Brace Issues:** Skin irritation, poor fit; requires regular brace adjustment.
- **Residual Deformity:** May require further treatment or surgery.

14. MPS - Classification

- ❖ Mucopolysaccharidosis (MPS) is a group of metabolic disorders caused by the absence or malfunctioning of lysosomal enzymes needed to break down molecules called glycosaminoglycans (GAGs)
- ❖ The accumulation of GAGs in cells, blood, and connective tissues leads to progressive cellular damage affecting appearance, physical abilities, organ and system functioning, and, in most cases, mental development
- ❖ MPS disorders are complex and vary widely in their presentation and severity
- ❖ Understanding the specific type and its implications is crucial for appropriate management and genetic counseling
- ❖ Advances in treatment and supportive care continue to improve the quality of life for individuals with MPS.

Table: Classification of Mucopolysaccharidosis (MPS)

MPS Type	Name	Deficient Enzyme	Primary Accumulated Substance	Key Clinical Features
MPS I AR	Hurler, Hurler-Scheie, Scheie Syndrome	α -L-iduronidase	Dermatan sulfate, Heparan sulfate	Skeletal abnormalities Developmental delay (severe in Hurler, mild in Scheie) Corneal clouding Cardiac issues
MPS II X-LR	Hunter Syndrome	Iduronate-2-sulfatase	Dermatan sulfate, Heparan sulfate	Mild to severe mental retardation Aggressive behavior Hearing loss Joint stiffness
MPS III AR	Sanfilippo Syndrome (A, B, C, D)	III-A - Heparan N-sulfatase III-B - α -N-acetylglucosaminidase	Heparan sulfate	Severe neurological symptoms Behavioral problems Sleep disturbances Less severe somatic symptoms

		III-C - Acetyl-CoAlpha-glucosaminide acetyltransferase III-D - N-Acetylglucosamine 6-sulfatase		
MPS IV AR	Morquio Syndrome (A, B)	Galactosamine-6-sulfatase (IV-A) Beta-galactosidase (IV-B)	Keratan sulfate	Skeletal dysplasia Short stature Corneal clouding Normal intelligence
MPS VI AR	Maroteaux-Lamy Syndrome	Arylsulfatase B	Dermatan sulfate	Similar to Hurler, but with normal intelligence Growth retardation Corneal clouding Cardiac issues
MPS VII AR	Sly Syndrome	β -Glucuronidase	Dermatan sulfate, Heparan sulfate, Chondroitin sulfate	Wide spectrum of severity Skeletal abnormalities Developmental delay Corneal clouding
MPS IX AR	Hyaluronidase Deficiency	Hyaluronidase	Hyaluronic acid	Periarticular soft tissue masses Short stature Mild skeletal abnormalities

- **Severity and Progression:** The severity and progression of symptoms can vary significantly, even within the same type.
- **Diagnosis:** Diagnosis often involves urine tests for GAGs, enzyme activity tests, and genetic testing.
- **Treatment:** Enzyme replacement therapy is available for some types, and supportive care is important for managing symptoms and improving quality of life.
- **Research:** Ongoing research includes gene therapy and other novel treatments.

15. Dog bite - guideline of management

Pre-Exposure Prophylaxis for Rabies

Intramuscular (IM) Regimen

- **Schedule:** 3 doses of rabies vaccine on Day 0, Day 7, and Day 21 or 28.

- **Vaccine Used:** HDCV (Human Diploid Cell Vaccine) or PCECV (Purified Chick Embryo Cell Vaccine)
- **Dosage:** 1.0 mL per dose
- **Route:** Intramuscular, usually in the deltoid region

Intradermal (ID) Regimen

- **Schedule:** 3 doses on Day 0, Day 7, and Day 21 or 28.
- **Vaccine Used:** HDCV or PCECV
- **Dosage:** 0.1 mL per dose
- **Route:** Intradermal, usually in the deltoid region

Post-Exposure Prophylaxis for Rabies

WHO Category I Exposure

- **Treatment:** No treatment required
- **Rationale:** Touching or feeding animals, licks on intact skin

WHO Category II Exposure

- **Treatment:** Immediate wound cleaning and vaccination
- **Schedule:** 3 doses on Day 0, Day 3, and Day 7
- **Vaccine Used:** HDCV or PCECV
- **Dosage:** 1.0 mL per dose (IM) or 0.1 mL per dose (ID)
- **Route:** Intramuscular or Intradermal

WHO Category III Exposure

- **Treatment:** Immediate wound cleaning, vaccination, and Rabies Immunoglobulin (RIG)
- **Schedule for Vaccine:** 5 doses on Day 0, Day 3, Day 7, Day 14, and Day 28
- **Vaccine Used:** HDCV or PCECV
- **Dosage for Vaccine:** 1.0 mL per dose (IM) or 0.1 mL per dose (ID)
- **Route for Vaccine:** Intramuscular or Intradermal
- **Rabies Immunoglobulin:** HRIG (Human Rabies Immunoglobulin) or ERIG (Equine Rabies Immunoglobulin)
- **Dosage for RIG:** 20 IU/kg (HRIG) or 40 IU/kg (ERIG)

- **Route for RIG:** Infiltration around the wound

Immunocompromised Individuals

- **Treatment:** Immediate wound cleaning, vaccination, and Rabies Immunoglobulin (RIG)
- **Schedule for Vaccine:** 5 doses on Day 0, Day 3, Day 7, Day 14, and Day 28, plus one additional dose on Day 90
- **Vaccine Used:** HDCV or PCECV
- **Dosage for Vaccine:** 1.0 mL per dose
- **Route for Vaccine:** Intramuscular
- **Rabies Immunoglobulin:** HRIG or ERIG
- **Dosage for RIG:** 20 IU/kg (HRIG) or 40 IU/kg (ERIG)
- **Route for RIG:** Infiltration around the wound

Schedule Type	Dosing Schedule	Vaccine Type	Route	Advantages
Pre-Exposure IM	Day 0, Day 7, Day 21 or 28	HDCV, PCECV	Intramuscular	- Long-lasting immunity - Suitable for travelers and those at high risk
Pre-Exposure ID	Day 0, Day 7, Day 21 or 28	HDCV, PCECV	Intradermal	- Cost-effective - Less vaccine volume required
Post-Exposure IM (Essen)	Day 0, Day 3, Day 7, Day 14, Day 28	HDCV, PCECV	Intramuscular	- Well-studied - Suitable for general population
Post-Exposure ID (Thai Red Cross)	Day 0 (2 doses), Day 3, Day 7, Day 28	HDCV, PCECV	Intradermal	- Cost-effective - Rapid induction of immunity
Post-Exposure IM (Zagreb)	Day 0 (2 doses), Day 7, Day 21	HDCV, PCECV	Intramuscular	- Shorter regimen - Less clinic visits required
Post-Exposure ID (Updated Thai)	Day 0, Day 3, Day 7	HDCV, PCECV	Intradermal	- Fewer doses - Cost-effective

Immunocompromised IM	Day 0, Day 3, Day 7, Day 14, Day 28, Day 90	HDCV, PCECV	Intramuscular	- Enhanced protection for immunocompromised individuals
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16. Enzyme replacement therapy

- **Mechanism of Action:** Direct supplementation of the deficient enzyme to restore metabolic function.
- **Indications:** Lysosomal storage diseases like Gaucher's disease, Fabry disease, Pompe disease, and Mucopolysaccharidosis.
- **Administration:** Intravenous infusion, sometimes intrathecal for CNS involvement.
- **Efficacy:** Effective in reducing substrate accumulation and ameliorating symptoms but may not reverse all disease manifestations.
- **Limitations:**
 - High cost
 - Lifelong treatment
 - Risk of immune reactions
 - Limited CNS penetration
- **Ethical and Social Considerations:** Accessibility and affordability, especially in resource-limited settings.
- **Recent Advances:** Development of more stable and effective recombinant enzymes, and exploration of oral enzyme formulations.

• Enzyme Replacement Therapies for Various Disorders

Generic Name	Enzyme Involved	Disease Targeted	Brand Name
<i>Alpha1-Proteinase inhibitor</i>	Alpha1-Antitrypsin	A1AT Deficiency	Prolastin-C, Glassia
<i>Alglucerase alfa</i>	β -Glucocerebrosidase	Gaucher	Ceredase*

<i>Imiglucerase</i>	β -Glucocerebrosidase	Gaucher	Cerezyme
<i>Taliglucerase alfa</i>	β -Glucocerebrosidase	Gaucher	Elelyso
<i>Velaglucerase alfa</i>	β -Glucocerebrosidase	Gaucher	VPRIV
<i>Pegademase</i>	Adenosine Deaminase	ADA Deficiency	Adagen
<i>Agalsidase beta</i>	Alpha-Galactosidase A	Fabry	Fabrazyme
<i>Alglucosidase alfa</i>	Acid alpha-Glucosidase	Pompe	Lumizyme
<i>Laronidase</i>	α -L-Iduronidase	Hurler, MPS I	Aldurazyme
<i>Idursulfase</i>	Iduronate-2-Sulfatase	Hunter, MPS II	Elaprase
<i>Elosulfase alfa</i>	N-Acetylgalactosamine-6 Sulfatase	Morquio Syndrome A, MPS IVA	Vimizim
<i>Galsulfase</i>	N-Acetylgalactosamine-4 Sulfatase	Maroteaux-Lamy, MPS VI	Naglazyme
<i>Sebelipase alfa</i>	Lysosomal Acid Lipase	Wolman, LAL Deficiency	Kanuma

- **Note:** Ceredase has been withdrawn from the market.

17. Monoclonal antibody in pediatric practice

- ❖ Monoclonal antibodies (mAbs) have increasingly become a vital part of therapeutic strategies in pediatric practice, addressing a range of conditions from autoimmune disorders to cancers
- ❖ Monoclonal antibodies represent a significant advancement in pediatric therapeutics, offering targeted treatments for a variety of serious conditions
- ❖ Their use requires careful consideration of the balance between therapeutic benefits and potential risks, as well as ongoing monitoring and patient education
- ❖ As research progresses, the scope of mAbs in pediatric practice is likely to expand further.

Table: Monoclonal Antibodies in Pediatric Practice

Monoclonal Antibody	Indication	Mechanism of Action	Notable Considerations
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Infliximab	Crohn's Disease Ulcerative Colitis Juvenile Idiopathic Arthritis	TNF- α inhibitor	Risk of serious infections TB screening required
Adalimumab	Juvenile Idiopathic Arthritis Crohn's Disease	TNF- α inhibitor	Self-administered via injection Similar considerations as Infliximab
Rituximab	B-cell Non-Hodgkin Lymphoma Autoimmune diseases (e.g., lupus nephritis)	CD20-directed cytolytic antibody	Infusion reactions Hepatitis B reactivation risk
Omalizumab	Severe Asthma Chronic Urticaria	Inhibits IgE binding	Administered every 2-4 weeks Anaphylaxis risk
Palivizumab	Prevention of RSV in high-risk infants	RSV F protein inhibitor	Seasonal administration during RSV season
Bevacizumab	Certain types of cancer (off-label in pediatrics)	VEGF inhibitor	Potential for impaired wound healing Risk of GI perforation
Eculizumab	Atypical Hemolytic Uremic Syndrome Paroxysmal Nocturnal Hemoglobinuria	Complement C5 inhibitor	Risk of meningococcal infections Vaccination required
Denosumab	Osteoporosis (in conditions like osteogenesis imperfecta)	RANKL inhibitor	Rarely used in pediatrics Risk of hypocalcemia
Natalizumab	Multiple Sclerosis (rare in pediatrics)	Integrin inhibitor	Risk of PML (progressive multifocal leukoencephalopathy)
Dupilumab	Moderate-to-severe Atopic Dermatitis	IL-4R α antagonist	Recently approved for adolescents Injection site reactions

- **Age Considerations:** Some mAbs are approved for use in certain pediatric age groups, while others are used off-label.
- **Monitoring:** Regular monitoring for side effects and efficacy is essential.
- **Infection Risk:** Many mAbs increase the risk of infections; prophylactic measures and screening are often necessary.
- **Cost and Access:** mAbs can be expensive and may require insurance approval or trial based sponsorship.
- **Emerging Therapies:** New mAbs are continually being developed and tested in clinical trials.

18. Plasmapheresis – indications

- ❖ Plasmapheresis, also known as therapeutic plasma exchange (TPE), is a procedure in which plasma is separated from the blood cells and replaced with a replacement solution, typically albumin or plasma
- ❖ This process is used to remove pathogenic substances from the blood, such as autoantibodies, immune complexes, or toxins
- ❖ Plasmapheresis has a wide range of indications in pediatric practice, particularly in autoimmune and other systemic disorders

Indications for Plasmapheresis in Pediatrics

1. *Autoimmune Neurological Disorders:*

- Acute inflammatory demyelinating polyneuropathy (Guillain-Barré syndrome).
- Chronic inflammatory demyelinating polyneuropathy (CIDP).
- Myasthenia gravis.
- Multiple sclerosis (acute fulminant).

2. *Renal Disorders:*

- Hemolytic uremic syndrome (atypical).
- Anti-GBM disease (Goodpasture's syndrome).
- Lupus nephritis with significant renal involvement.
- ANCA-associated vasculitis with severe renal involvement.

3. *Hematological Disorders:*

- Thrombotic thrombocytopenic purpura (TTP).
- Hemolytic disease of the newborn (as adjunct to phototherapy and/or exchange transfusion).
- Sickle cell disease (acute chest syndrome or multi-organ failure).

4. *Metabolic Disorders:*

- Familial hypercholesterolemia (particularly in cases resistant to conventional therapy).
- Refractory hyperlipidemia.

5. **Rheumatologic Disorders:**

- Juvenile dermatomyositis with severe disease.
- Refractory systemic lupus erythematosus (SLE).

6. **Transplantation:**

- ABO-incompatible organ transplantation (pre-transplant).
- Antibody-mediated rejection of transplanted organs.

7. **Toxicology:**

- Severe drug overdose or poisoning where the drug is amenable to removal by plasmapheresis (e.g., certain medications, toxins).

8. **Miscellaneous:**

- Severe refractory dermatologic conditions (e.g., pemphigus vulgaris).
- Certain cases of severe sepsis or multi-organ failure where conventional treatments have failed.

Additional Considerations

- **Patient Selection:** Careful patient selection is crucial, as plasmapheresis is an invasive procedure with potential complications.
- **Timing and Frequency:** The timing and frequency of plasmapheresis sessions depend on the underlying condition and clinical response.
- **Replacement Fluids:** Choice between fresh frozen plasma and albumin based on clinical scenario.
- **Monitoring:** Close monitoring during and after the procedure for potential complications like hypotension, electrolyte imbalances, and allergic reactions.
- **Combination with Other Therapies:** Often used in conjunction with pharmacotherapy for optimal outcomes.

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19. Newborn screening - tests and interpretations

- **Metabolic Disorders:** These are conditions that affect the body's metabolism. Examples include:

- Phenylketonuria (PKU)
- Maple syrup urine disease (MSUD)
- Medium-chain acyl-CoA dehydrogenase deficiency (MCADD)
- Very long-chain acyl-CoA dehydrogenase deficiency (VLCADD)
- Isovaleric acidemia (IVA)
- **Endocrine Disorders:** These affect hormone-producing glands. Examples include:
 - Congenital hypothyroidism
 - Congenital adrenal hyperplasia (CAH)
- **Hemoglobinopathies:** These are blood disorders. Examples include:
 - Sickle cell disease
 - Beta-thalassemia
 - Hemoglobin C disease
- **Cystic Fibrosis:** A genetic disorder that affects the respiratory and digestive systems.
- **Severe Combined Immunodeficiency (SCID):** A group of disorders causing a malfunction in the humoral and cell mediated immune system.
- **Hearing Loss:** Early detection can allow for interventions that support language development.
- **Critical Congenital Heart Disease (CCHD):** Heart defects that can cause serious, life-threatening symptoms; especially duct dependant lesions need early intervention.
- **Organic Acidemias:** Disorders in the metabolism of amino acids. Examples include:
 - Propionic acidemia (PA)
 - Methylmalonic acidemia (MMA)
- **Fatty Acid Oxidation Disorders:** Problems with breaking down fats for energy. Examples include:
 - Long-chain hydroxyacyl-CoA dehydrogenase deficiency (LCHADD)
 - Trifunctional protein deficiency (TFP)

- **Amino Acid Disorders:** Conditions affecting the metabolism of amino acids. Examples include:
 - Tyrosinemia type I
 - Argininosuccinic aciduria
- **Urea Cycle Disorders:** These affect the body's ability to remove ammonia from the bloodstream. Examples include:
 - OTC deficiency
 - Citrullinemia
 - Argininemia
- **Lysosomal Storage Disorders:** Conditions caused by the malfunction of lysosomal enzymes. Examples include:
 - Krabbe disease
 - Pompe disease
 - Gaucher disease
- **Peroxisomal Disorders:** Such as X-linked adrenoleukodystrophy.
- **Biotinidase Deficiency:** A condition where the body can't recycle biotin (vitamin B7).
- **Galactosemia:** A condition where the body can't process galactose, associated with E.coli sepsis and hypoglycemia.
- **Spinal Muscular Atrophy (SMA):** A genetic disorder that affects muscle control, presents as floppy infant.

How Newborn Screening is Done:

- **Blood Spot Screening:** A few drops of blood are taken from the newborn's heel, usually within 24-48 hours after birth, and placed on a special filter paper card. This blood is then tested for various genetic, endocrine, metabolic, and hemoglobin disorders.
- **Hearing Screening:** Two different tests are commonly used to screen newborns' hearing. The otoacoustic emissions (OAE) test measures sound waves produced in the inner ear, and the auditory brainstem response (ABR) test measures how the auditory nerve and brain respond to sound.

- **Pulse Oximetry:** This is a non-invasive test that estimates the percentage of hemoglobin in blood that is saturated with oxygen. It's used to screen for CCHD.
- **Confirmatory Testing:** If a screening result is positive, further testing is done to confirm the diagnosis. This may include repeat blood tests, urine tests, imaging studies, or specialized genetic testing.

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20. Temper tantrum presentation and management

- ❖ Temper tantrums are a normal part of child development, typically seen in toddlers and preschool-aged children
- ❖ They are often expressions of frustration or a desire for attention and can manifest as crying, screaming, kicking, hitting, or holding their breath
- ❖ Managing temper tantrums effectively is crucial for both the child's development and the caregiver's well-being
- ❖ Temper tantrums are a challenging but normal part of early childhood development
- ❖ Effective management involves a combination of prevention, calm and consistent responses during tantrums, and teaching appropriate emotional regulation skills
- ❖ Consistency across caregivers and environments is key
- ❖ If tantrums are severe, frequent, or persist beyond the expected age range, professional evaluation may be warranted to rule out underlying issues.

Presentation of Temper Tantrums

1. **Age of Onset:** Commonly begin around 18 months and peak between 2 and 3 years.
2. **Triggers:** Often triggered by frustration, fatigue, hunger, or being denied something.
3. **Behavioral Manifestations:**
 - Crying, screaming, or yelling.
 - Physical actions like kicking, hitting, or throwing objects.
 - Breath-holding spells in some cases.

- Refusal to comply with requests or instructions.
- May become more intense if the child is tired, hungry, or stressed.

Management Strategies

1. Prevention:

- Establish a routine to provide a sense of security.
- Set clear, consistent limits and expectations.
- Use positive reinforcement to encourage good behavior.
- Identify triggers and try to avoid or mitigate them.

2. During a Tantrum:

- Stay calm and do not react with anger or frustration.
- Ensure the child is in a safe environment where they can't hurt themselves or others.
- Offer comfort or distraction, if effective.
- Avoid giving in to unreasonable demands made during tantrums.
- Use time-outs judiciously, ensuring the child understands why it's happening.

3. Post-Tantrum:

- Once calm, discuss the behavior with the child in simple terms.
- Teach and model appropriate ways to express feelings.
- Reinforce the idea that while it's okay to feel upset, certain behaviors are not acceptable.

4. Communication:

- Encourage the use of words to express feelings as language skills develop.
- Validate their feelings but guide them towards appropriate expressions.

5. Consistency:

- Apply rules and consequences consistently.
- All caregivers should respond to tantrums in a similar manner to avoid confusion.

6. **Professional Consultation:**

- If tantrums are excessively frequent, intense, or continue beyond the typical age, consider consulting a pediatrician or child psychologist.
- Evaluation for underlying developmental, behavioral, or emotional issues may be necessary.

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